CHAPTER 16

Strabismus

Introduction

Definitions

Normal ocular alignment involves parallelism of the visual axes for distance vision, or intersection of the visual axes at the point of fixation for near.

1. **Strabismus** (squint) is a misalignment of the eyes.
2. **Orthophoria** implies perfect ocular alignment *without effort*, even in the absence of any stimulus for fusion. It is rare; most individuals manifest mild heterophoria.
3. **Heterophoria** ('phoria) implies a tendency of the eyes to deviate (latent squint). Ocular alignment is maintained with effort.
4. **Heterotropia** ('tropia) implies a manifest squint—the eyes are misaligned. A phoria may become a tropia if:
   - Muscle strength is inadequate to maintain ocular alignment.
   - Stimulus for fusion is weak (e.g. uniocular blurred vision).
   - Neurological pathways subserving ocular coordination break down.
5. **Prefixes** 'eso' and 'exo' imply inward and outward ocular deviations respectively. For example: an exophoria is a tendency of the eyes to diverge, while esotropia is a manifest convergent squint. Ocular misalignment may also be vertical, where the prefixes hypo- (downward) and hyper- (upward) apply, or torsional.
6. **Visual axis** (line of vision) passes from the fovea, through the nodal point of the eye to the point of fixation (object of regard). The two visual axes normally intersect at the point of fixation. The fovea is usually slightly temporal to the posterior pole (geometrical centre at the back of the globe); the visual axis therefore cuts the cornea slightly nasal of centre.
7. **Anatomical axis** is a line passing from the posterior pole through the centre of the cornea.
8. **Angle kappa** is the angle subtended by the visual and anatomical axes and is usually about 5° (Fig. 16.1). Angle kappa is positive when the fovea is temporal to the posterior pole, and negative when the converse applies. Abnormalities of angle kappa may result in pseudo-strabismus (*see* later).

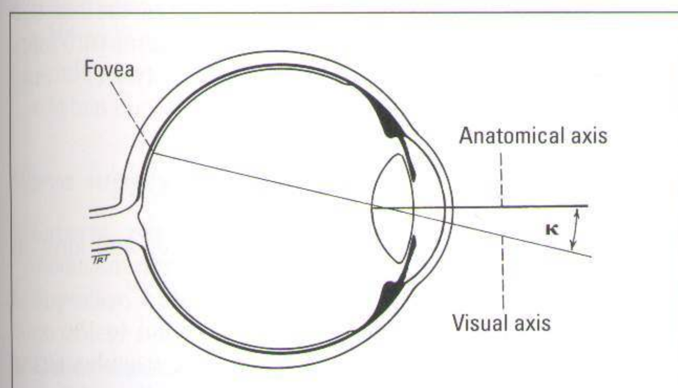


Fig. 16.1
Angle kappa

Anatomy of extraocular muscles

General principles

The lateral and medial orbital walls are at an angle of 45° to each other (Fig. 16.2a). The orbital axis therefore forms an angle of 22.5° with both lateral and medial walls. For the sake of simplicity this angle is usually regarded as being 23°. When the eye is looking straight ahead at a fixed point on the horizon with the head erect (primary position of gaze), the visual axis forms an angle of 23° with the orbital axis (Fig. 16.2b). The actions of the extraocular muscles depend on the position of the globe at the time of muscle contraction.

1. **Primary action** of a muscle is its major effect when the eye is in the primary position.
2. **Subsidiary actions** are the additional effects on the position of the eye.
3. **Listing plane** is an imaginary coronal plane passing through the centre of rotation of the globe. The globe rotates on the axes of Fick, which intersect in Listing plane (Fig. 16.3).
   - The globe rotates left and right on the vertical Z-axis.
   - The globe moves up and down on the horizontal X-axis.

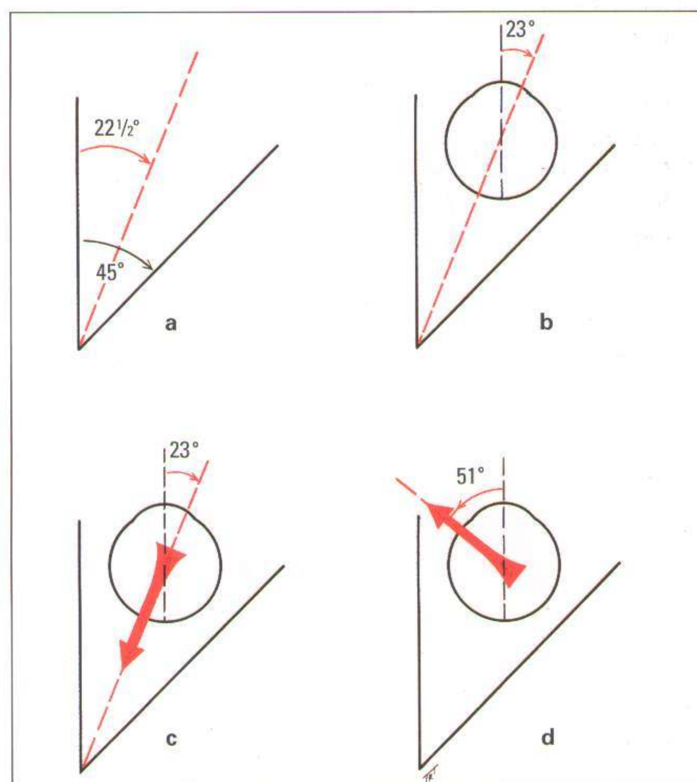


Fig. 16.2
Anatomy of extraocular muscles (*see text*)

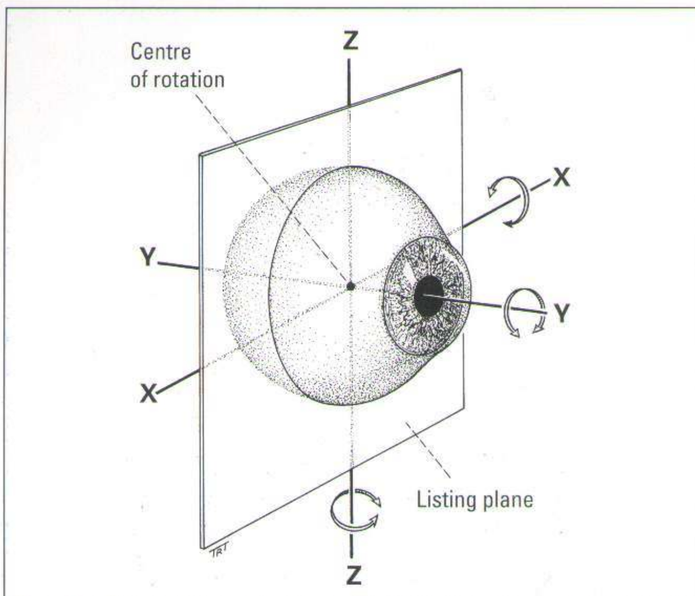


Fig. 16.3
Listing plane and axes of Fick (*see text*)

- Torsional movements occur on the *Y*-axis which traverses the globe from front to back (similar to the anatomical axis of the eye).

Horizontal rectus muscles

When the eye is in the primary position, the horizontal recti are purely horizontal movers on the vertical Z-axis and have only primary actions.

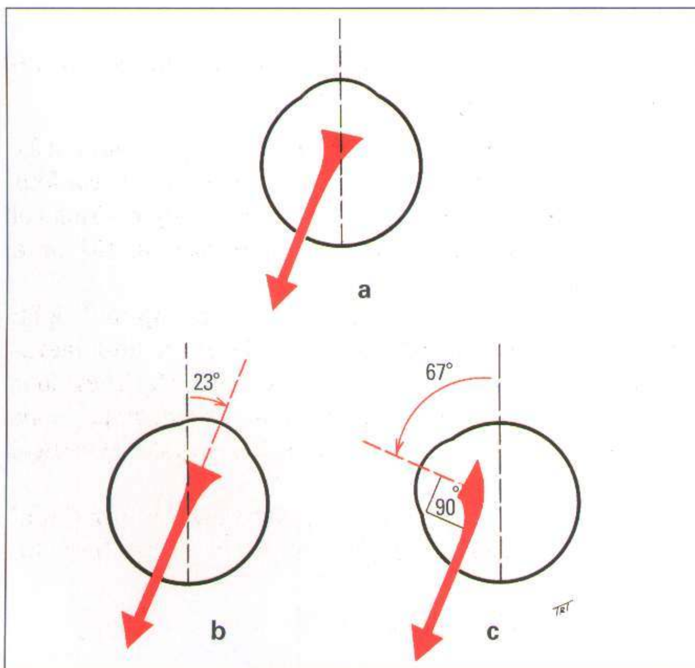


Fig. 16.4
Actions of the right superior rectus muscle

1. **Medial rectus** originates at the annulus of Zinn at the orbital apex and inserts 5.5 mm behind the nasal limbus. Its sole action is adduction.
2. **Lateral rectus** originates at the annulus of Zinn and inserts 6.9 mm behind the temporal limbus. Its sole action is abduction.

Vertical rectus muscles

The vertical recti run in line with the orbital axis and are inserted in front of the equator. They therefore form an angle of 23° with the visual axis (*see* Fig. 16.2c).

1. **Superior rectus** originates from the upper part of the annulus of Zinn and inserts 7.7 mm behind the superior limbus.
   - The primary action of the superior rectus is elevation (Fig. 16.4a). Secondary actions are adduction and intorsion.
   - When the globe is abducted 23°, the visual and orbital axes coincide (Fig. 16.4b). In this position it has no subsidiary actions and can only act as an elevator. This is therefore the optimal position of the globe for testing the function of the superior rectus muscle.
   - If the globe were adducted 67°, the angle between the visual and orbital axes would be 90° (Fig. 16.4c). In this position the superior rectus could only act as an intortor.
2. **Inferior rectus** originates at the lower part of the annulus of Zinn and inserts 6.5 mm behind the inferior limbus.
   - The primary action of the inferior rectus is depression. Secondary actions are adduction and extorsion.
   - When the globe is abducted 23°, the inferior rectus acts purely as a depressor. As for superior rectus, this is the optimal position of the globe for testing the function of the inferior rectus muscle.
   - If the globe were adducted 67°, the inferior rectus could only act as an extortor.

Spiral of Tillaux

This is an imaginary line joining the insertions of the four recti and is an important anatomical landmark when performing surgery. The insertions get further away from the limbus and make a spiral pattern. The medial rectus insertion is closest (5.5 mm), followed by the inferior rectus (6.5 mm), lateral rectus (6.9 mm) and superior rectus (7.7 mm) (Fig. 16.5).

Oblique muscles

The obliques are inserted behind the equator and form an angle of 51° with the visual axis (*see* Fig. 16.2d).

1. **Superior oblique** originates superomedial to the optic foramen. It passes forwards through the trochlea at the angle between the superior and medial walls and is then

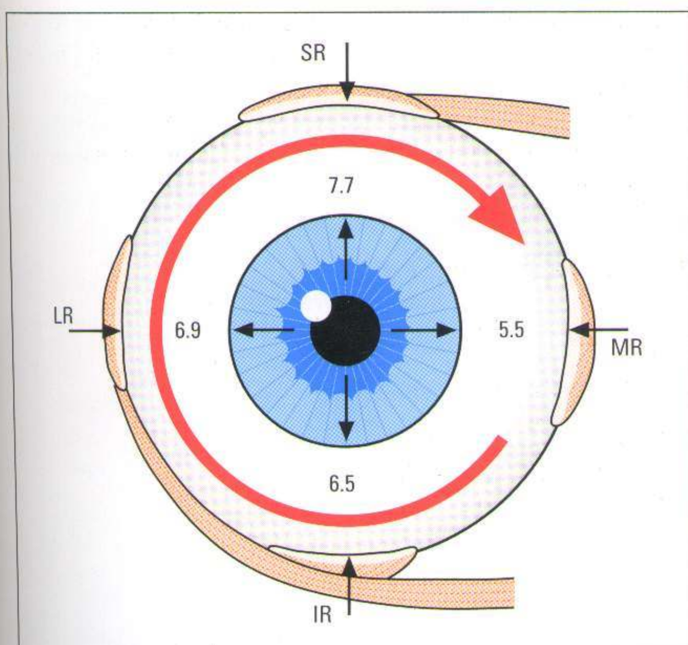


Fig. 16.5
Spiral of Tillaux (*see text*)

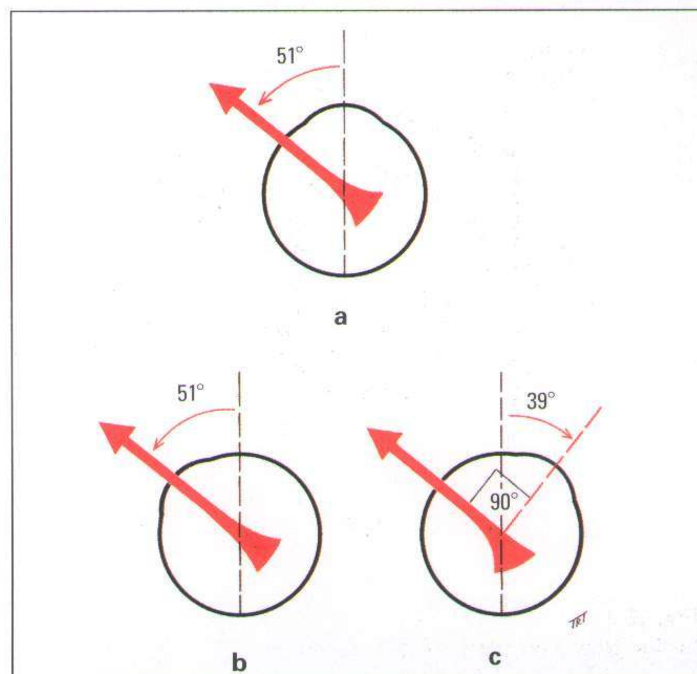


Fig. 16.6
Actions of the right superior oblique muscle (*see text*)

reflected backwards and laterally to insert in the posterior upper temporal quadrant of the globe.

- The primary action of the superior oblique is intorsion (Fig. 16.6a) Secondary actions are depression and abduction.
- When the globe is adducted 51°, the visual axis coincides with the line of pull of the muscle (Fig. 16.6b). In this position it can only act as a depressor. This is therefore the best position of the globe for testing the action of the superior oblique muscle.
- When the eye is abducted 39°, the visual axis and the superior oblique make an angle of 90° with each other (Fig. 16.6c). In this position the superior oblique can only cause intorsion.

2. **Inferior oblique** originates from a small depression just behind the orbital rim lateral to the lacrimal sac. It passes backwards and laterally, to insert in the posterior lower temporal quadrant of the globe, *close to the macula*.
   - The primary action of the inferior oblique is extorsion. Secondary actions are elevation and abduction.
   - When the globe is adducted 51°, the inferior oblique acts only as an elevator.
   - When the eye is abducted 39°, its main action is extorsion.

Nerve supply

1. **Lateral rectus** is supplied by the 6th cranial nerve (abducent nerve—abducting muscle).
2. **Superior oblique** is supplied by the 4th cranial nerve (trochlear nerve—muscle associated with the trochlea).
3. **Other muscles** and the levator muscle of the upper lid are supplied by the 3rd (oculomotor) nerve.

Ocular movements

Ductions

These are monocular movements around the axes of Fick. They consist of adduction, abduction, elevation, depression, intorsion and extorsion. They are tested by occluding the fellow eye and asking the patient to follow a target in each direction of gaze.

Versions

These are binocular, simultaneous, conjugate movements (in the same direction) (Fig. 16.7, top).

- Dextroversion and laevoversion (gaze right; gaze left), elevation (upgaze) and depression (downgaze). These four movements bring the globe into the *secondary* positions of gaze by rotation around either a horizontal (X) or a vertical (Z) axis of Fick.
- Dextroelevation and dextrodepression (gaze up and right; gaze down and right) and laevoelevation and laevodepression (gaze up and left; gaze down and left). These four oblique movements bring the eyes into the *tertiary* positions of gaze by rotation around both the horizontal and vertical axes.
- Dextrocycloversion and laevocycloversion (torsional movement of the superior limbi of both eyes to the right; torsion to the left).

Vergences

These are binocular, simultaneous, disjunctive movements (in opposite directions) (Fig. 16.7, bottom). Convergence is simul-

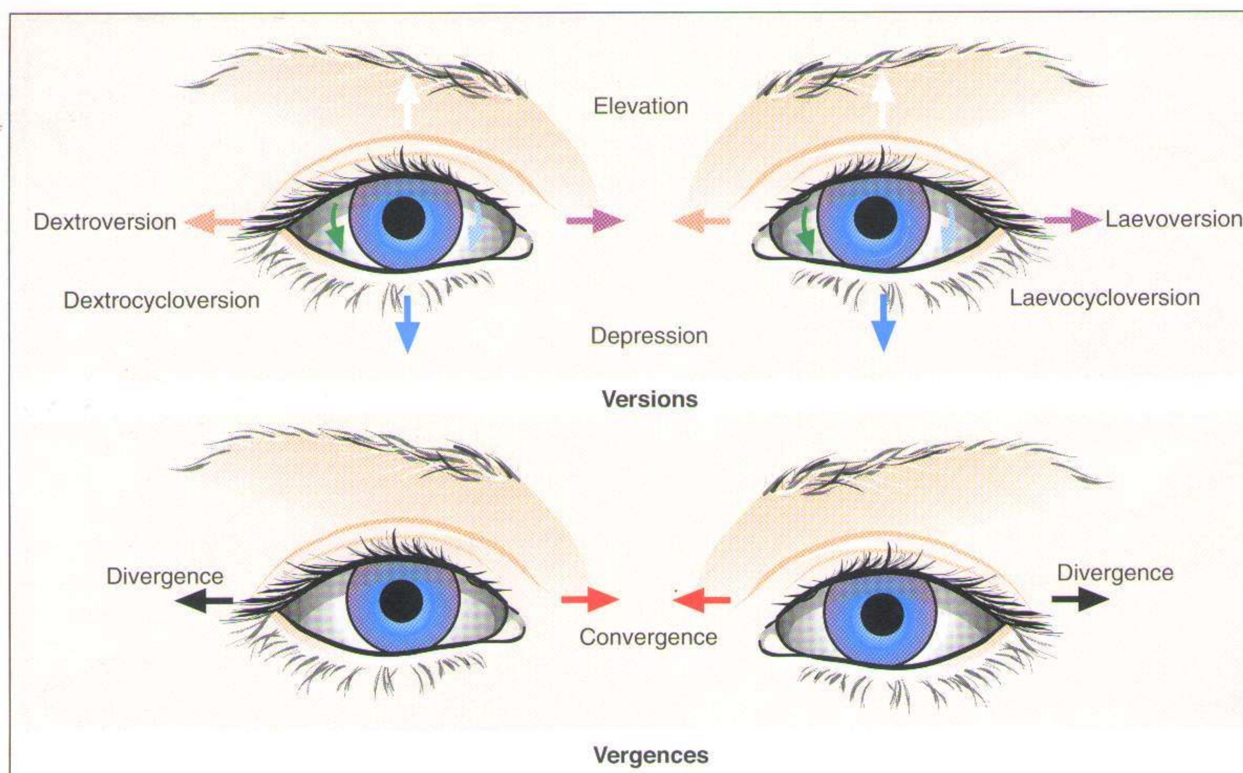


Fig. 16.7 Binocular movements (Courtesy of Wilmer Institute)

taneous adduction (inward turning); divergence is turning outwards from a convergent position. Convergence may be voluntary or reflex. Reflex convergence has four components:

1. **Tonic** convergence, which implies inherent innervational tone to the medial recti, when the patient is awake.
2. **Proximal** convergence is induced by psychological awareness of a near object.
3. **Fusional** convergence is an optomotor reflex, which maintains binocular single vision (BSV), by ensuring that similar images are projected onto corresponding retinal areas of each eye. It occurs without a change in refractive state of the eye and is initiated by bi-temporal retinal image disparity.
4. **Accommodative** convergence is induced by the act of accommodation as part of the synkinetic-near reflex. Each dioptre of accommodation is accompanied by a constant increment in accommodative convergence, giving the '*accommodative convergence by accommodation*' (AC/A) ratio. This is the amount of convergence in prism dioptres (Δ) per dioptre (D) change in accommodation. The normal value is 3–5 Δ. This means that 1 D of accommodation is associated with 3–5 Δ of accommodative convergence. It will be shown later that abnormalities of the AC/A ratio play an important role in the aetiology of strabismus.

Positions of gaze

1. **Six cardinal positions** of gaze are those in which one muscle in each eye has moved the eye into that position as follows:
   - Dextroversion (right lateral rectus and left medial rectus).
   - Laevoversion (left lateral rectus and right medial rectus).
   - Dextroelevation (right superior rectus and left inferior oblique).
   - Laevoelevation (left superior rectus and right inferior oblique).
   - Dextrodepression (right inferior rectus and left superior oblique).
   - Laevodepression (left inferior rectus and right superior oblique).
2. **Nine diagnostic positions of gaze** are those in which deviations are measured. They consist of the six cardinal positions, the primary position, elevation and depression (Fig. 16.8).

Laws of ocular motility

1. **Agonist–antagonist** pairs are muscles of the *same* eye that move the eye in *opposite* directions. The *agonist* is the primary muscle moving the eye in a given direction. The *antagonist* acts in the opposite direction to the agonist. For example, the right lateral rectus is the antagonist to the right medial rectus.
2. **Synergists** are muscles of the *same* eye that move the eye in the *same* direction. For example, the right superior rectus and right inferior oblique act synergistically in elevation.
3. **Yoke muscles** are *pairs of muscles*, one in each eye, that produce conjugate ocular movements. For example, the yoke muscle of the left superior oblique is the right inferior rectus.
4. **Sherrington law** of reciprocal innervation (inhibition) states that increased innervation to an extraocular muscle (e.g. right medial rectus) is accompanied by a reciprocal decrease in innervation to its antagonist (e.g. right lateral

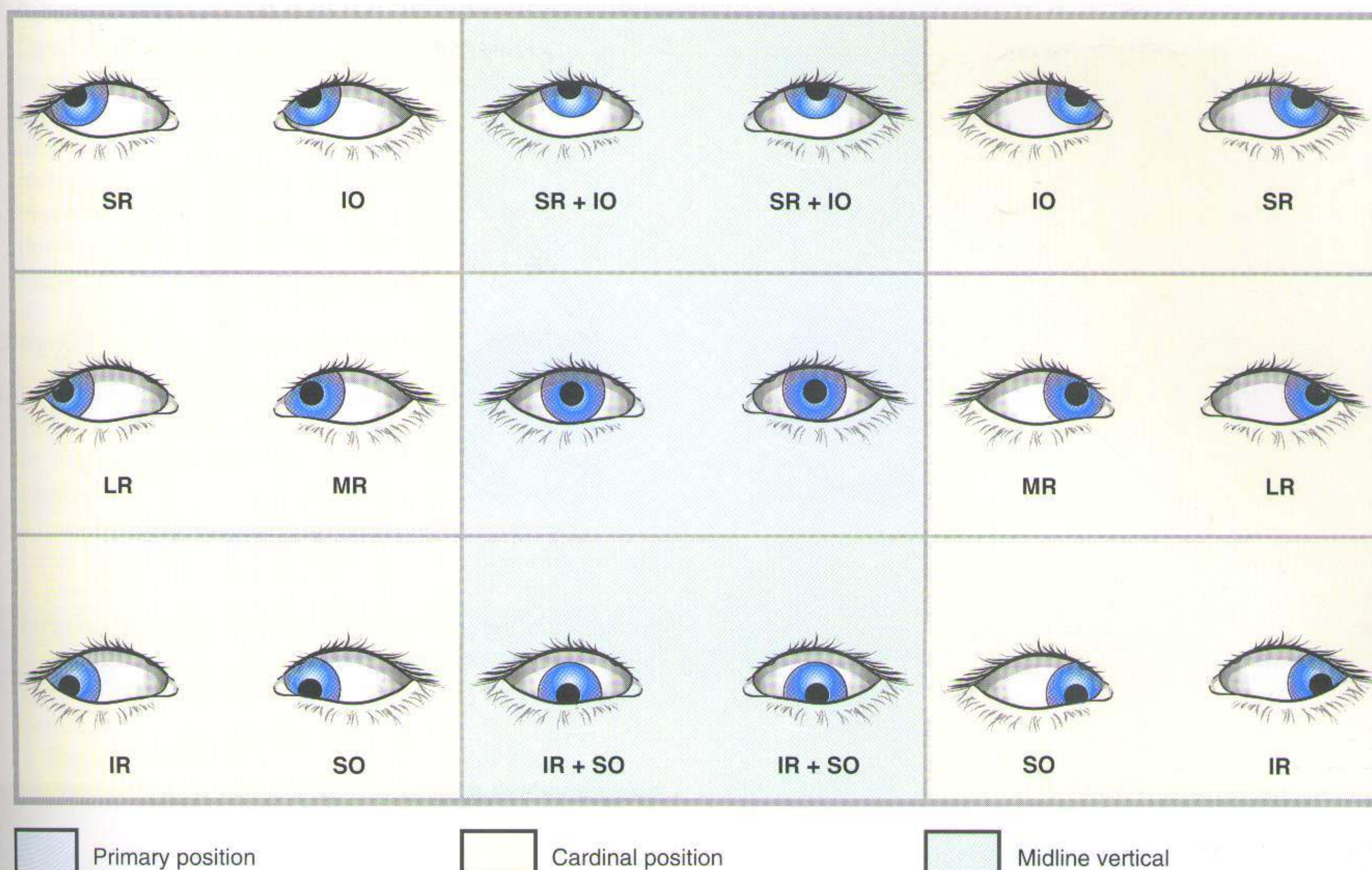


Fig. 16.8
Diagnostic positions of gaze (*see text*)

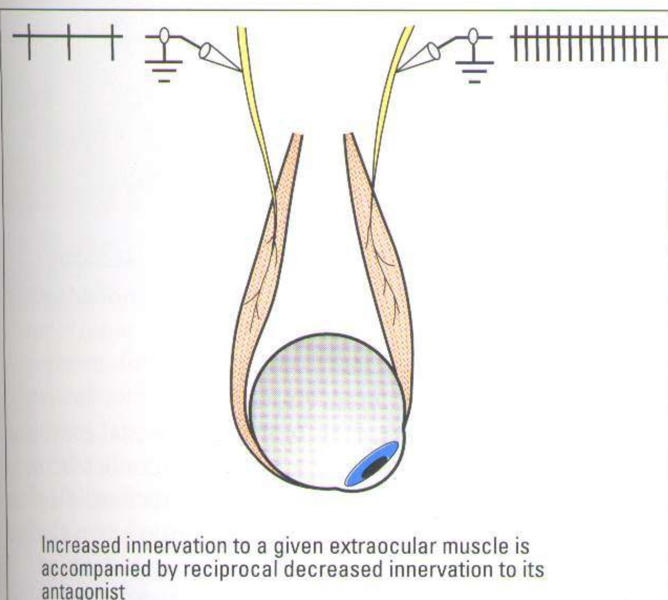


Fig. 16.9
Sherrington law of reciprocal innervation

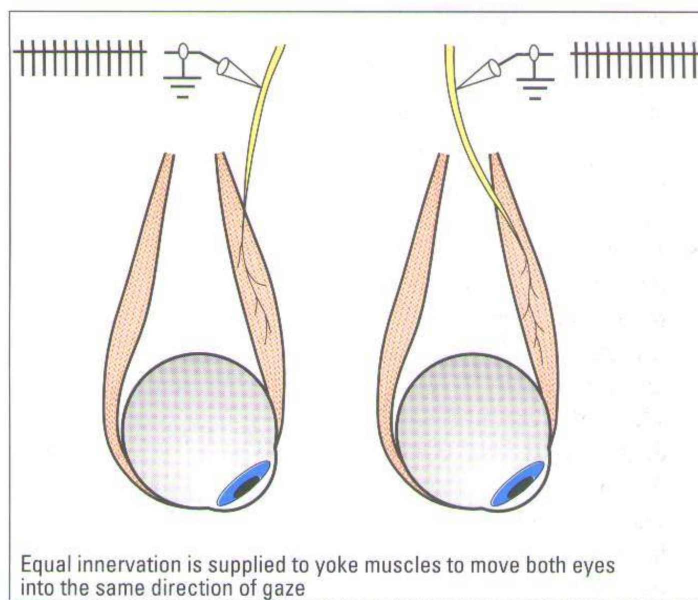


Fig. 16.10
Hering law of equal innervation of yoke muscles

rectus) (Fig. 16.9). This means that when the medial rectus contracts the lateral rectus automatically relaxes and vice versa. Sherrington law applies to both versions and vergences.

5. **Hering law** of equal innervation states that during any conjugate eye movement, equal and simultaneous innervation flows to the yoke muscles (Fig. 16.10). In the case of a paretic squint, the amount of innervation to both

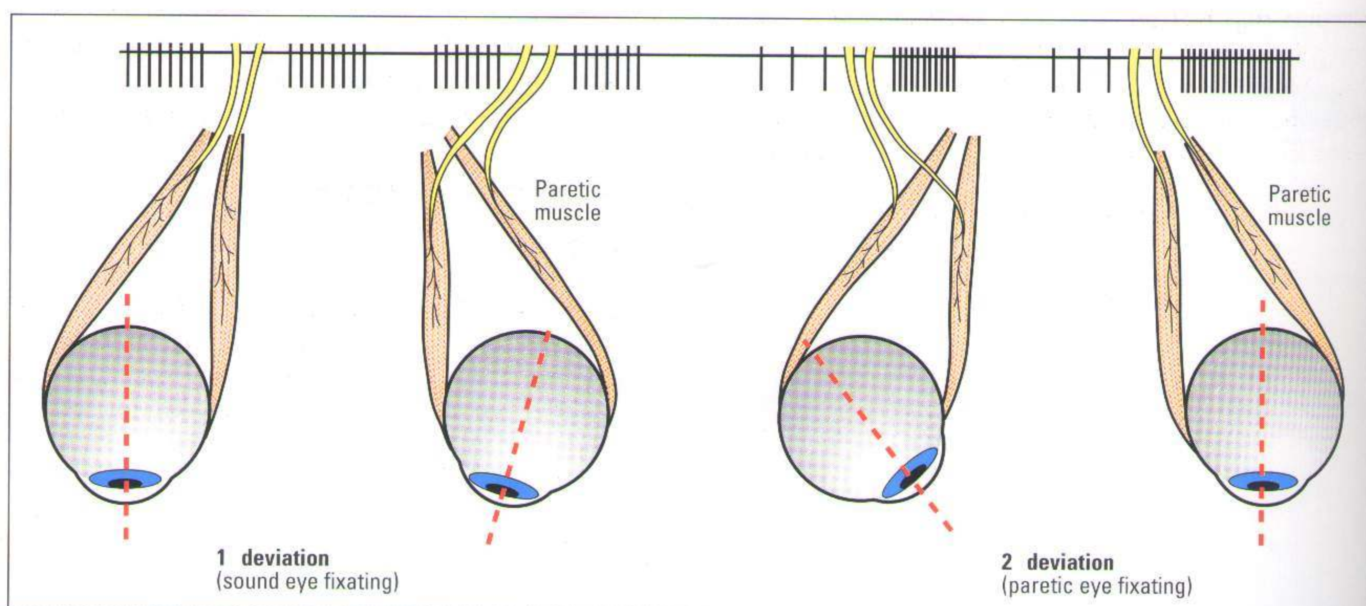


Fig. 16.11
Primary and secondary deviations in paretic strabismus (*see text*)

eyes is symmetrical, and always determined by the fixating eye, so that the angle of deviation will vary according to which eye is used for fixation. For example if, in the case of a left lateral rectus palsy, the right normal eye is used for fixation, there will be an inward deviation of the left eye due to the unopposed action of the antagonist of the paretic left lateral rectus (left medial rectus). The amount of misalignment of the two eyes in this situation is called the *primary deviation* (Figs 16.11, left and 16.12a). If the paretic left eye is now used for fixation, additional innervation will flow to the left lateral rectus, in order to establish this. However, according to Hering law, an equal amount of innervation will also flow to the right medial rectus (yoke muscle). This will result in an overaction of the right medial rectus and an excessive amount of adduction of the right eye. The amount of misalignment between the two eyes in this situation is called the *secondary deviation* (Figs 16.11, right and 16.12b). In a paretic squint, the secondary deviation exceeds the primary deviation.

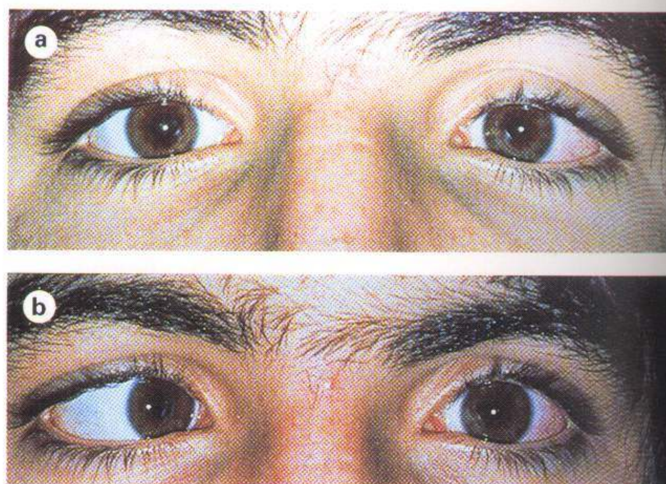


Fig. 16.12
Left sixth nerve palsy. **(a)** Primary deviation; **(b)** secondary deviation (Courtesy of Wilmer Institute)

Functional consequences of strabismus

Amblyopia

Classification

Amblyopia is the unilateral, or (rarely) bilateral, decrease of best-corrected visual acuity caused by form vision deprivation and/or abnormal binocular interaction, for which there is no pathology of the eye or visual pathway.

1. **Strabismic** amblyopia results from abnormal binocular interaction where there is continued monocular suppression of the deviating eye. It is characterized by an impairment of vision which is present even when the eye is forced to fixate.
2. **Anisometropic** amblyopia is caused by a difference in refractive error of even as little as 1.0 D sphere. It results from abnormal binocular interaction from the superimposition of focused and unfocused images or from the superimposition of large and small images (aniseikonia). There may also be an element of form vision deprivation as one eye constantly receives a blurred image. It is frequently associated with microstrabismus and may coexist with strabismic amblyopia.

3. **Stimulus deprivation** amblyopia results from form vision deprivation. It may be unilateral or bilateral and is caused by opacities in the media (e.g. cataract) or severe ptosis.
4. **Iso-ametropic** amblyopia results from form vision deprivation. It is bilateral and is caused by high symmetrical refractive errors, usually hypermetropia.
5. **Meridional** amblyopia results from form vision deprivation in one meridian. It can be unilateral or bilateral and is caused by uncorrected astigmatism.

Diagnosis

1. **Visual acuity**. In the absence of an organic lesion, a difference in best corrected visual acuity of two lines or more is indicative of amblyopia. Visual acuity in amblyopia is often better while reading single letters than a row of letters. This 'crowding' phenomenon occurs to a certain extent in normal individuals but is more profound in amblyopes.
2. **Neutral density filter** is useful to differentiate impaired vision due to organic disease from amblyopia. A neutral density filter, which reduces visual acuity by two lines in a normal eye, is used as follows:
   a. Best corrected visual acuity is determined.
   b. The filter is placed in front of the eye and visual acuity measured.
   c. No significant drop in visual acuity implies amblyopia.
   d. A significant drop in visual acuity implies an organic lesion.
3. **Grating acuity** (ability to perceive a grid pattern of different frequencies) often exceeds spatial (Snellen) acuity in amblyopia.

> *NB:* Visual fields and colour vision are normal.

Treatment

The sensitive period during which amblyopia can be reversed is up to 7–8 years in strabismic amblyopia and longer (up to 11–12 years) for anisometropic amblyopia.

1. **Occlusion** of the normal eye, to encourage use of the amblyopic eye, is the most effective treatment. The regimen, full-time or part-time, depends on the age of the patient and the density of amblyopia. The younger the patient, the more rapid the improvement, although the greater the risk of inducing amblyopia in the normal eye. It is therefore very important to monitor visual acuity in both eyes during treatment. The better the visual acuity at the start of occlusion, the shorter the duration required. If there has been no improvement after 6 months of occlusion, further treatment is unlikely to be fruitful.
2. **Penalization**, in which vision in the normal eye is blurred with atropine, is an alternative method. It may be used in the treatment of relatively mild amblyopia (6/24 or better) in association with hypermetropia. Penalization does not work as quickly as occlusion, and is only effective if vision in the normal eye is made worse that the vision in the amblyopic eye, at least for near.

> *NB:* It is essential to exclude organic disease prior to commencing treatment for amblyopia.

Confusion and diplopia

Basic concepts

1. **Binocular single vision** (BSV) involves the simultaneous use of both eyes with bifoveal fixation, so that each eye contributes to a common single perception of the object of regard (Fig. 16.13a). Conditions necessary for BSV are:
   - Overlap of visual fields.
   - Accurate neuromuscular development and co-ordination, so that the visual axes are directed at the object.
   - Normal visual pathways.
   - Approximately equal image clarity and size in both eyes.
   - Corresponding retinal areas so that the eyes are cyclopean.
2. **Confusion** is the simultaneous appreciation of two superimposed but dissimilar images caused by stimulation of corresponding points (usually the foveae) by images of different objects (Fig. 16.13b).
3. **Diplopia** is the simultaneous appreciation of two images of the same object. It results from images of the same object falling on non-corresponding

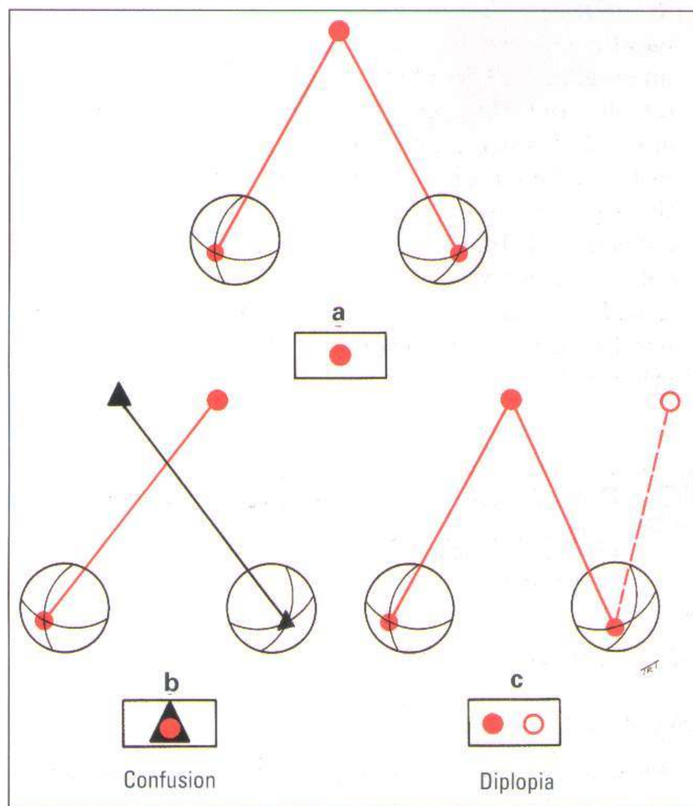


Fig. 16.13
Double vision (*see text*)

retinal points (Fig. 16.13c). Simultaneous (visual) perception is the ability to use both eyes simultaneously.

4. **Visual direction** is the projection of a given retinal element in a specific direction in subjective space.
   a. ***Principal*** visual direction is the direction in external space interpreted as the line of sight. This is normally the visual direction of the fovea.
   b. ***Secondary*** visual directions are the projecting directions of extrafoveal points with respect to the principal direction of the fovea.
5. **Projection** is the interpretation of the position of an object in space on the basis of stimulated retinal elements.
   - If a red object stimulates the right fovea (F), and a black object which lies in the nasal field stimulates a temporal retinal element (T), the red object will be interpreted by the brain as having originated from the straight ahead position and the black object will be interpreted as having originated in the nasal field (Fig. 16.14a). Similarly, nasal retinal elements project into the temporal field, upper retinal elements into the lower field and vice versa.
   - With both eyes open, the red fixation object is now stimulating both foveae, which are corresponding retinal points. The black object is now not only stimulating the temporal retinal elements in the right eye but also the nasal elements of the left eye. The right eye therefore projects the object into its nasal field and the left eye projects the object into its temporal field. However, because both of these retinal elements are corresponding points, they will both project the object into the same position in space (the left side) and there will be no double vision.

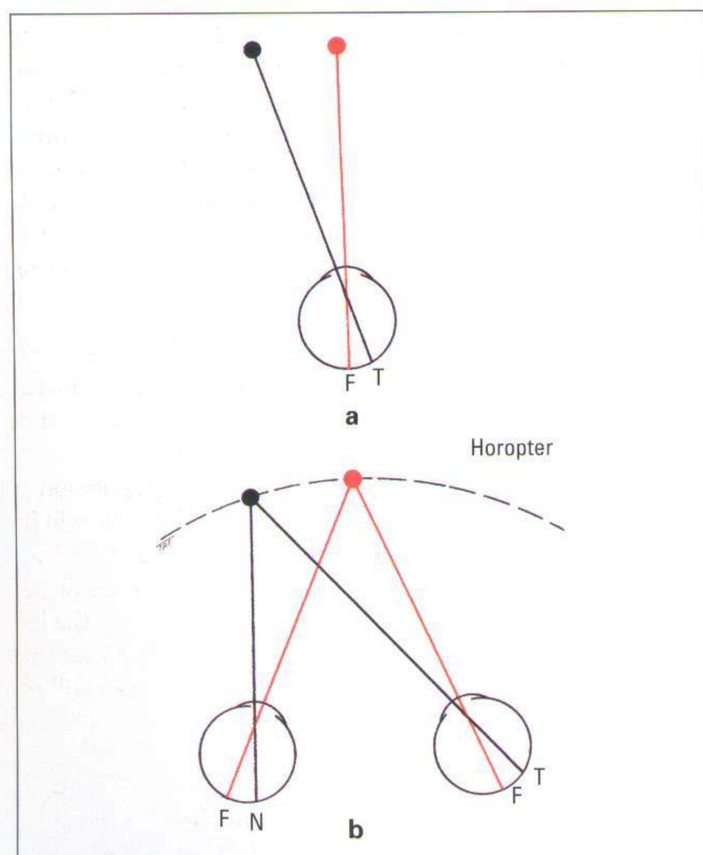


Fig. 16.14
Principles of projection (*see text*)

6. **Retino-motor values**. The image of an object in the peripheral visual field falls on an extrafoveal element. To establish fixation on this object a saccadic version of accurate amplitude is required. Each extrafoveal retinal element therefore has a retino-motor value proportional to its distance from the fovea, which guides the amplitude of saccadic movements required to 'look at it'. Retino-motor value, zero at the fovea, increases progressively towards the retinal periphery.
7. **Corresponding points** are areas on each retina that share the same subjective visual direction (for example, the foveae, which project straight ahead). Points on the nasal retina of one eye have corresponding points on the temporal retina of the other eye. This is the basis of normal retinal correspondence. For example, an object producing images on the right nasal retina and the left temporal retina will be projected into the right side of visual space.
8. **Horopter** is an imaginary plane in external space, all points on which stimulate corresponding retinal elements and are therefore seen singly (Fig. 16.14b). This plane passes through the intersection of the visual axes and therefore includes the point of fixation in BSV.
9. **Panum fusional area of BSV** is a zone in front of and behind the horopter in which objects (although not accurately stimulating corresponding retinal elements) are seen singly. Objects outside Panum area appear double. This is the basis of physiological diplopia. Panum area is narrow at fixation (6 seconds of arc) and broadens towards the periphery. Therefore objects on the horopter are seen singly. Objects in Panum fusional area are seen singly and stereoscopically. Objects outside Panum fusional area appear double.
10. **Sensory** fusion involves the integration, by the visual areas of the cerebral cortex, of two similar images, one from each eye, into one image. It may be central, which integrates images falling on the foveae, or peripheral, which integrates images falling outside the foveae.
11. **Motor** fusion involves the maintenance of physical alignment of the eyes to sustain bifoveal fixation. It is driven by retinal image disparity, which stimulates fusional vergences.
12. **Fusional vergence** involves disjunctive eye movements to overcome retinal image disparity. Fusional vergence amplitudes can be measured with prisms or on the synoptophore. Normal values are:
    - Convergence: about 15 Δ for distance and 25 Δ for near.

- Divergence: about 6 Δ for distance and 12 Δ for near.
- Vertical: 2–3 Δ.
- Cyclovergence: about 2–3°.

Fusional convergence helps to control an exophoria whereas fusional divergence helps to control an esophoria. The fusional vergence mechanism may be decreased by fatigue or illness, converting a phoria to a tropia. The amplitude of fusional vergence mechanisms can be improved by orthoptic exercises, particularly that of near fusional convergence for the relief of convergence insufficiency.

13. **Stereopsis** is the perception of depth (the third dimension, the first two being height and width). It arises when objects behind and in front of the point of fixation (but within Panum fusional area) stimulate horizontally disparate retinal elements simultaneously. The fusion of such disparate images results in a single visual impression perceived in depth. A solid object is seen stereoscopically (in 3D) because each eye sees a slightly different aspect of the object.

Sensory adaptation to strabismus

The ocular sensory system in children has the ability to adapt to anomalous states (confusion and diplopia) by two mechanisms: (a) *suppression* and (b) *abnormal retinal correspondence* (ARC). They occur because of the plasticity of the developing visual system in children under the age of 6–8 years. Rarely adults who develop sudden-onset strabismus are able to ignore the second image and therefore do not complain of diplopia.

1. **Suppression** involves active inhibition, by the visual cortex, of an image from one eye when *both eyes are open*. Stimuli for suppression include diplopia, confusion and a blurred image resulting from astigmatism/anisometropia. Clinically, suppression may be:
   a. ***Central*** or ***peripheral***. In central suppression the image from the fovea of the deviating eye is inhibited to avoid confusion. Diplopia, on the other hand, is eradicated by the process of peripheral suppression, in which the image from the peripheral retina of the deviating eye is inhibited.
   b. ***Monocular*** or ***alternating***. Suppression is monocular when the image from the dominant eye always predominates over the image from the deviating (or more ametropic) eye, so that the image from the latter is constantly suppressed. This type of suppression leads to the development of amblyopia. When suppression alternates (switches from one eye to the other), amblyopia does not develop.
   c. ***Facultative*** or ***obligatory***. Facultative suppression occurs only when the eyes are misaligned. Obligatory suppression is present at all times, irrespective of whether the eyes are deviated or straight.
2. **Abnormal retinal correspondence** (ARC) is a condition in which non-corresponding retinal elements acquire a common subjective visual direction. The fovea of the fixating eye is thus paired with a non-foveal element of the deviated eye. ARC is a positive sensory adaptation to strabismus (as opposed to suppression), which allows some binocular vision with limited fusion to be maintained in the presence of a heterotropia. ARC is most frequently present in small angle esotropia. It is rare in accommodative esotropia because of the variability of the angle of deviation, or in large angle deviations because the separation of the images is too great. It is also rarely found in exotropia because the deviation is often intermittent. When a child first develops strabismus, the following events occur:
   a. The fovea of the squinting eye is suppressed to avoid confusion.
   b. Diplopia will occur, since non-corresponding retinal elements receive the same image.
   c. To avoid diplopia, the patient will develop either peripheral suppression of the squinting eye or ARC.
   d. If suppression occurs this will subsequently lead to strabismic amblyopia.

> ***NB:*** The disadvantage of strongly entrenched ARC is that following surgery the patient may not revert to normal retinal correspondence. Therefore the angle of deviation may return to the pre-surgical state in an attempt to regain binocular vision.

Motor adaptation to strabismus

This involves the adoption of an abnormal head posture and occurs in adults who cannot suppress or in children with good binocular vision potential. Under strabismic conditions, patients will adopt an abnormal head posture to maintain BSV and eliminate diplopia. The patient will turn the head *into* the direction of the field of action of the weak muscle, so that the eyes are then automatically turned in the opposite direction and as far as possible *away* from its field of action (i.e. the head will turn where the eye cannot).

1. **Horizontal** deviation will result in a face turn. For example, if one of the muscles that turn the eyes to the left is paralysed, the face will also be turned to the left so that the eyes no longer need to look towards the left.
2. **Vertical** deviation will result in elevation or depression of the chin. If one of the elevators is weak, the chin will be elevated so that the eyes become depressed (Fig. 16.15).
3. **Torsional** deviation will result in a head tilt to one or the other shoulder. For example, if an intortor such as the left superior oblique is paralysed, the left eye will become extorted. In order to compensate for this, the head will tilt to the right shoulder, effectively 'intorting' the left eye (Fig. 16.16).

> ***NB:*** As a rule of thumb, a head tilt often accompanies vertical misalignments. The tilt is usually towards the lower eye, but not due to the vertical misalignment, but to the accompanying (and less apparent) torsional deviation.



Fig. 16.15
Compensatory chin elevation and head tilt



Fig. 16.16
Compensatory head tilt

Clinical evaluation

History

1. **Age of onset** can give an indication as to the aetiology of a squint. The earlier the onset, the more probable the need for surgical correction. The later the onset, the greater the likelihood of an accommodative component. Inspection of previous photographs may be useful for the documentation of strabismus or an abnormal head posture.
2. **Variability** is significant because intermittent strabismus indicates some degree of binocularity. An alternating deviation suggests symmetrical visual acuity in both eyes.
3. **General health** or developmental problems are significant: (e.g. children with cerebral palsy have an increased incidence of strabismus).
4. **Birth history**, including period of gestation, birth weight and any problems with delivery or in utero.
5. **Family history** is important because strabismus is frequently familial, although there is no definitive inheritance pattern. It is also important to know what therapy was necessary in other family members.

Visual acuity

Definition

Spatial visual acuity is the ability to distinguish separate elements of a target and identify it as a whole. It is quantified by the minimum angle of separation (subtended at the nodal point of the eye) between two objects that allows them to be perceived as separate. The normal minimum angle of separation is 1 minute or less, and corresponds to letters on the 6/6 line of the Snellen chart, when viewed from a distance of 6 metres.

Testing in preverbal children

An estimate of the comparative vision between the two eyes can be gained from simple examination and observation of the child.

1. **Occlusion of one eye**, if strongly objected to by the child, indicates poorer acuity in the other eye (Fig. 16.17).

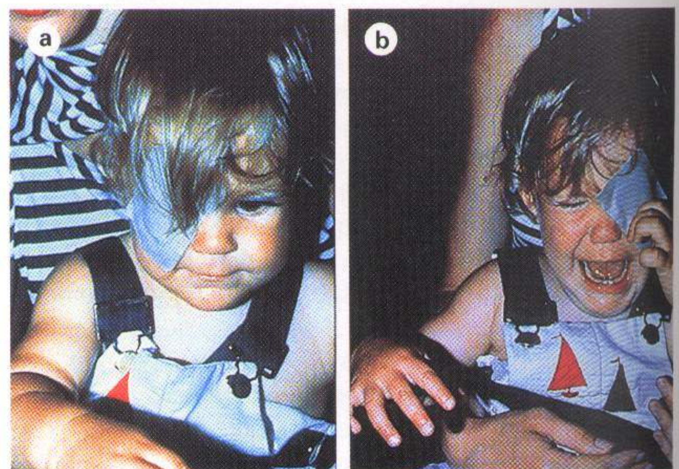


Fig. 16.17
Amblyopic right eye. (**a**) No objection to occlusion of amblyopic eye; (**b**) objection to occlusion of normal eye
(Courtesy of Wilmer Institute)

2. **Fixation test** is performed as follows:
   a. A 16 Δ base-down prism is placed over one eye and the other is occluded.
   b. The eye behind the prism is therefore forced to elevate, to take up fixation.
   c. The eye behind the prism is then observed.
   d. Fixation is then graded as central or non-central and steady or unsteady.
   e. The other eye is uncovered and the ability to maintain fixation is observed.
   f. If fixation immediately returns to the uncovered eye, then visual acuity is impaired.
   g. If fixation is maintained through a blink, then visual acuity is good.
   h. If the patient alternates fixation, then the two eyes have equal vision.
   i. The test is repeated with the prism over the other eye.
   j. Monocular fixation should be central, steady and maintained in each eye.
3. '**Hundreds and thousands**' sweet test is a gross test which is seldom performed (Fig. 16.18). In principle, if the child is able to see and pick up small sweets at 33 cm, visual acuity is at least 6/24.
4. **Rotation test** is a gross qualitative test of the ability of an infant to fixate with both eyes open. The test is performed as follows:
   a. The examiner holds the child facing him and rotates briskly through 360°.
   b. If vision is normal, the eyes will deviate in the direction of rotation under the influence of the vestibulo-ocular response. The eyes intermittently flick back to the primary position to produce a rotational nystagmus.
   c. When rotation stops, the nystagmus should also cease due to suppression of post-rotatory nystagmus by fixation.
   d. If vision is severely impaired, the induced nystagmus does not stop when rotation ceases because the vestibulo-ocular response is not blocked by visual feedback.
5. **Preferential looking** tests can be used from early infancy. They are based on the fact that infants prefer to look at a pattern rather than a homogeneous stimulus. The infant is exposed to a stimulus and the examiner observes the eyes for fixation movements. Two examples are Teller acuity cards, which consist of black stripes of varying thickness, and Cardiff acuity cards, which consist of shapes with variable outlines (Fig. 16.19). Low-frequency (thick) gratings or shapes with a bold outline are seen more easily than those with thin outlines, and an assessment of visual acuity is made accordingly. Since grating acuity often exceeds Snellen acuity in amblyopia, Teller cards may overestimate visual acuity.
6. **Pattern visually evoked potential** gives a representation of spatial acuity but is more regularly used in the diagnosis of optic neuropathy.
7. **Optokinetic nystagmus** may provide an estimation of visual acuity dependent on the size of the stripes used.

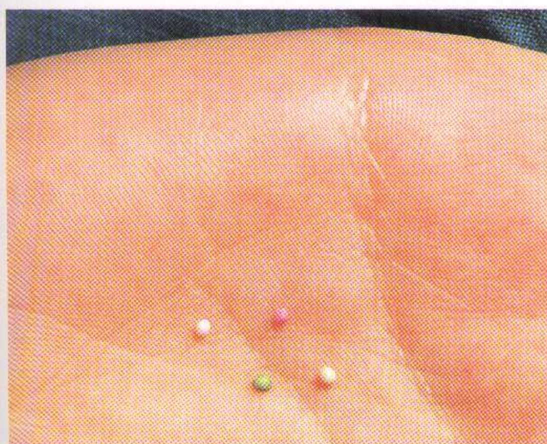
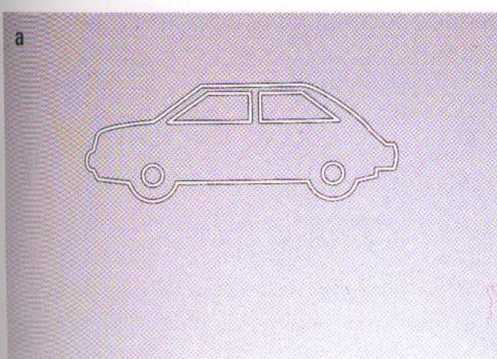


Fig. 16.18
'Hundreds and thousands' test (*see text*)

In verbal children

1. **At age 2 years**, most children will have sufficient language skills to undertake a picture-naming test such as the Kay pictures (Fig. 16.20).
2. **At age 3 years**, most children will be able to undertake the matching of single-letter optotypes as in the Sheridan–Gardiner test (Fig. 16.21). This test has the disadvantage of overestimating acuity in the amblyopic eye as it eliminates the crowding phenomenon. The Keeler LogMAR crowded test (Fig. 16.22) is more accurate in amblyopia as it requires the child to match one letter out of a group of optotypes, which relates more accurately to Snellen acuity.
3. **At age 4 years**, most children will be able to perform a linear Snellen acuity test.



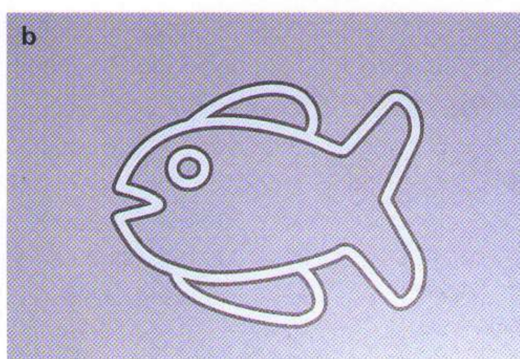


Fig. 16.19
Cardiff acuity cards (*see text*)

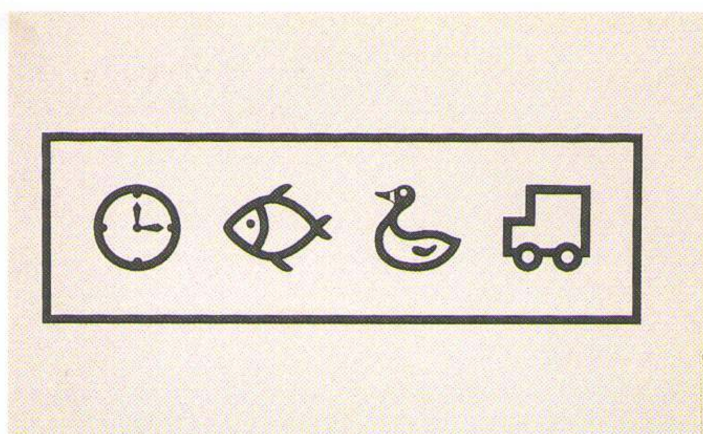


Fig. 16.20
Kay pictures (Courtesy of E. Dawson)



Fig. 16.21
Sheridan–Gardiner test (*see text*)



Fig. 16.22
Keeler LogMAR crowded test (*see text*) (Courtesy of E. Dawson)

Tests for stereopsis

Stereopsis is measured in seconds of arc (1° = 60 minutes of arc; 1 minute = 60 seconds of arc). It is useful to remember that normal spatial visual acuity is 1 minute and normal



Fig. 16.23
Titmus test (*see text*)

stereo-acuity is 60 seconds (which equals 1 minute). The lower the value, the better the acuity.

Titmus

This test consists of a three-dimensional polaroid vectograph consisting of two plates in the form of a booklet which are viewed through polaroid spectacles. On the right is a large fly, and on the left are a series of circles and animals (Fig. 16.23). The test is performed at a distance of 16 inches (405 mm).

1. **Fly** is a test of gross stereopsis (3000 seconds of arc) and is especially useful for young children. The fly should appear 'solid' and the child is encouraged to pick up one of its wings. In the absence of gross stereopsis the fly will appear as an ordinary flat photograph. If the book is inverted, the targets will appear to recede. If the patient states that the fly's wings are still 'popping out', then they are not appreciating stereoscopic vision.
2. **Circles** comprise a graded series which tests fine depth perception. Each of the nine squares contains four circles. One of the circles in each square has a degree of disparity and will appear forward of the plane of reference in the presence of normal stereopsis. The angle of stereopsis is calculated from a chart provided with the test. The degree of disparity ranges from 800 to 40 seconds of arc. If a

patient perceives the circle to be shifted off to the side, then they are not appreciating stereoscopic vision, but are using monocular clues instead.

3. **Animals** is similar to the circles test but consists of three rows of animals, one of which will appear forward of the plane of reference. The degree of disparity ranges from 400 to 100 seconds of arc.

TNO

This random dot test consists of seven plates, which are viewed with red–green spectacles. Each plate contains various shapes (squares, crosses etc.) created by random dots in complementary colours. Some shapes are visible even without red–green spectacles (Fig. 16.24a) while others are 'hidden', and only apparent to an individual with stereopsis, while wearing red–green spectacles (Fig. 16.24b). The first three plates are used to establish the presence of stereoscopic vision and subsequent plates to quantify it. Because there are no monocular clues, the TNO test provides a truer measurement of stereopsis than the Titmus test. The degree of disparity ranges from 480 to 15 seconds of arc.

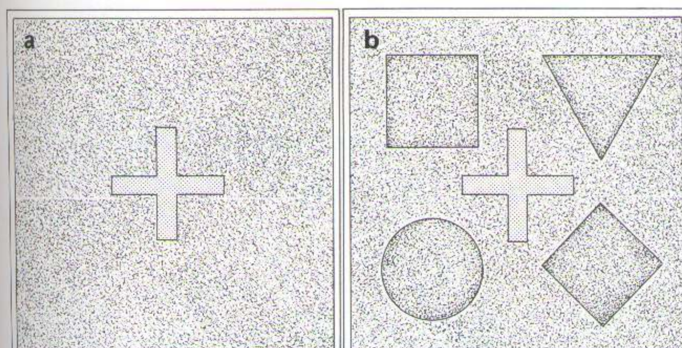


Fig. 16.24
TNO test (*see text*)

Lang

This test does not require special spectacles; the targets are seen alternately by each eye through the built-in cylindrical lens elements. Displacement of the dots creates disparity and the patient is asked to name or point to a simple shape, such as a star, on the card (Fig. 16.25). The Lang is especially useful in assessing stereopsis in very young children and babies, as they will instinctively reach out to touch the pictures. The examiner can also observe the child's eye movements from picture to picture on the card. The degree of disparity is quite gross, ranging from 1200 to 600 seconds of arc.

Frisby

This test consists of three transparent plastic plates of varying thickness. On the surface of each plate are printed four squares of small random shapes (Fig. 16.26). One of the squares contains a 'hidden' circle, in which the random shapes are printed on the reverse of the plate. The patient is required to identify this hidden circle. The test does not require special spectacles because the disparity is created by the thickness of the plate and can be varied by increasing or decreasing the working distance. The degree of disparity ranges from 600 to 15 seconds of arc.

Base-out prism

This is a quick and easy method for detecting BSV in children who cannot perform the stereo tests described above. The test is performed by placing a 20 Δ base-out prism in front of one eye (in this case the right). This displaces the retinal image temporally with resultant diplopia. The examiner observes corrective eye movements as follows:

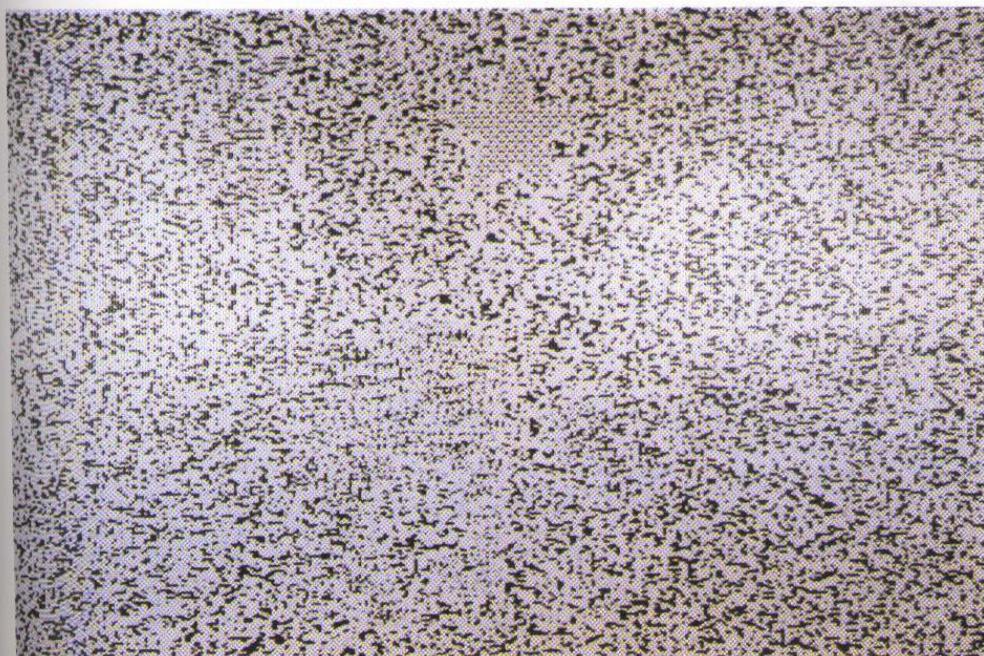


Fig. 16.25
Lang test (*see text*)

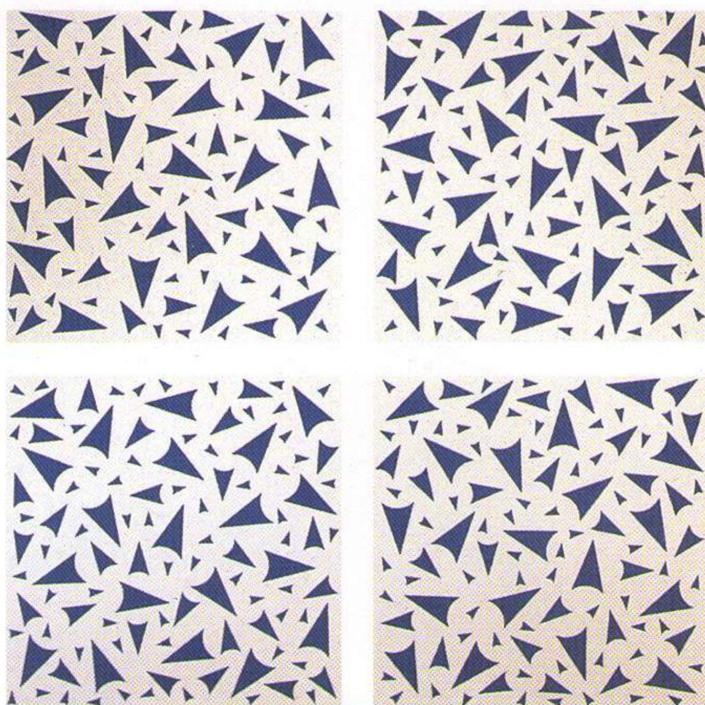


Fig. 16.26
Frisby test (*see text*)

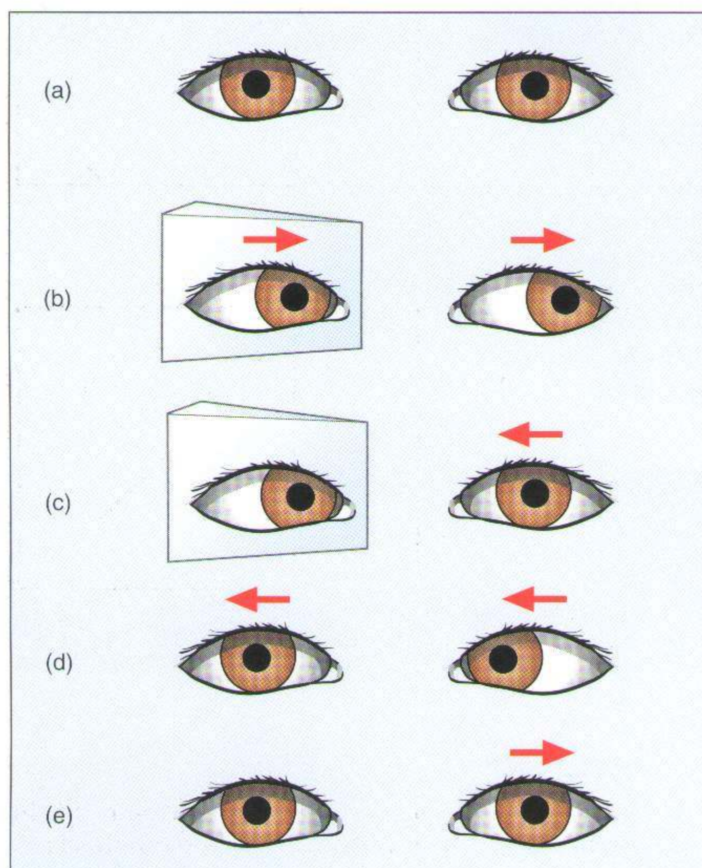


Fig. 16.27
Base-out prism test (*see text*)

a. There will be a shift of the right eye to the left to resume fixation (right adduction) with a corresponding shift of the left eye to the left (left abduction) in accordance with Hering law (Fig. 16.27b).



Fig. 16.28
Red–green goggles (Courtesy of Wilmer Institute)

b. The left eye will then make a corrective refixational saccade to the right (left re-adduction) (Fig. 16.27c).
c. On removal of the prism both eyes move to the right (Fig. 16.27d).
d. The left eye then makes an outward fusional movement (Fig. 16.27e).

> ***NB:*** Most children with good BSV should be able to overcome a 20 Δ prism; if not, weaker prisms (16 Δ or 12 Δ) may be tried.

Tests for sensory anomalies

Worth four-dot

1. Procedure

a. The patient wears a red lens in front of the right eye, which filters out all colours except red.
b. A green lens is placed in front of the left eye, which will filter out all colours except green (Fig. 16.28).
c. The patient then views a box with four lights: one red, two green and one white.

2. Results (Fig. 16.29)

- If all four lights are seen, normal fusion is present.
- If all four lights are seen in the presence of a manifest deviation, ARC is present.
- If two red lights are seen, left suppression is present.
- If three green lights are seen, right suppression is present.
- If two red and three green lights are seen, diplopia is present.
- If the green and red lights alternate, alternating suppression is present.

Bagolini striated glasses

Each lens is covered with fine striations which convert a point source of light into a line, similar to the Maddox rod (*see below*).

1. Procedure

a. The two lenses are placed at 45° and 135° in front of each eye and the patient fixates a punctate light source (Fig. 16.30).

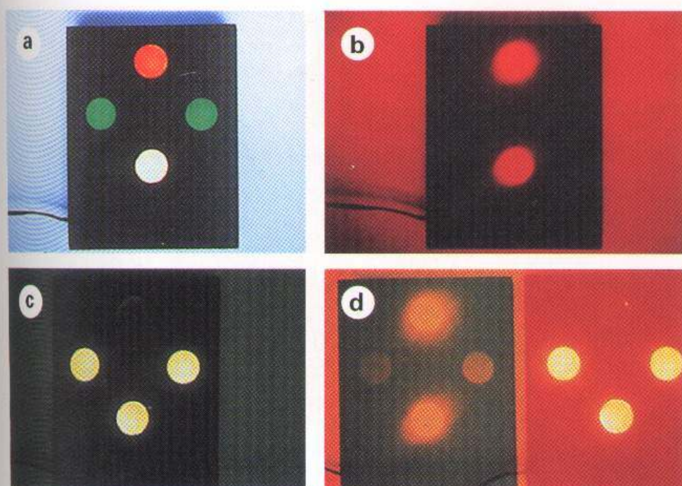
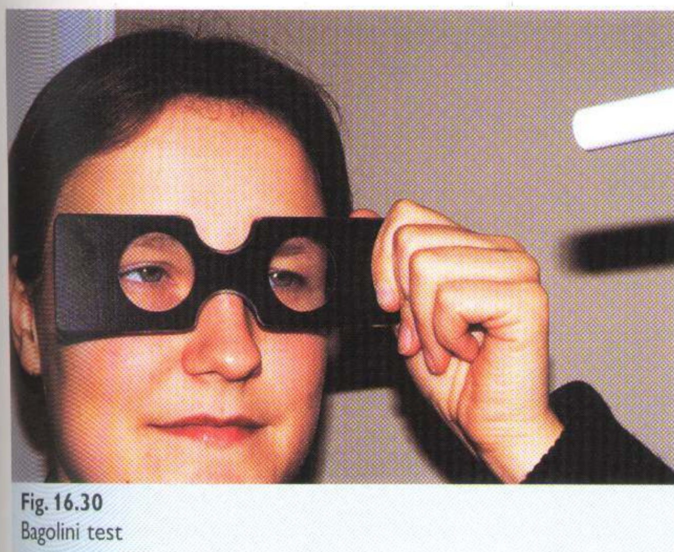


Fig. 16.29
Possible results of the Worth four-dot test (*see text*) (Courtesy of Wilmer Institute)


Fig. 16.30
Bagolini test

b. Each eye perceives an oblique line of light, perpendicular to that perceived by the fellow eye (Fig. 16.31).
c. Dissimilar images are thus presented to each eye under binocular viewing conditions.

2. **Results** cannot be interpreted correctly unless it is known whether or not strabismus is present:
   - If the two streaks intersect at their centres in the form of an oblique cross (an 'X'), the patient is either orthophoric or (if strabismic) has ARC (Fig. 16.32a).
   - If the two lines are seen but they do not form a cross, diplopia is present (Fig. 16.32b).
   - If only one streak is seen, there is no simultaneous perception (Fig. 16.32c).
   - If a small gap is seen in one of the streaks, a central suppression scotoma is present (Fig. 16.32d).

After-image

This test demonstrates the visual direction of the foveae.


Fig. 16.31
Appearance of point of light through Bagolini lenses (Courtesy of Wilmer Institute)

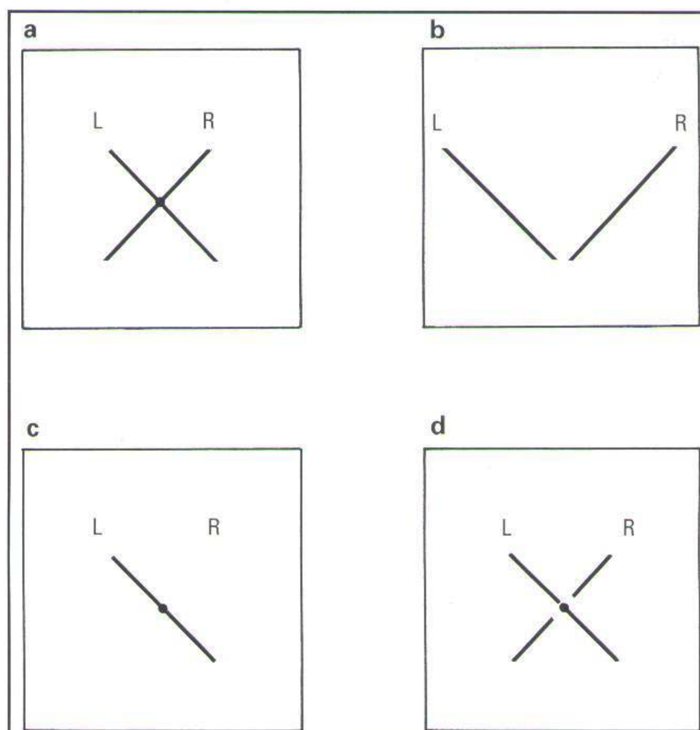
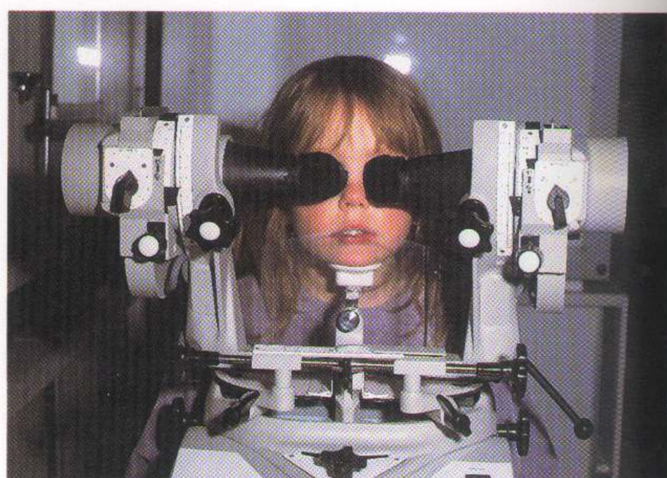


Fig. 16.32
Possible results of the Bagolini test (*see text*)

1. **Procedure**
   a. One fovea is stimulated by a vertical bright flash of light and the fellow fovea is stimulated by a horizontal flash of light (Fig. 16.33).
   b. The vertical flash of light is harder to suppress and should therefore be applied to the deviating eye.
2. **Results**. The patient then draws the relative positions of the after-images.
   - If the two after-images are seen as a cross, retinal correspondence is normal (Fig. 16.34a).
   - If the two images do not cross, ARC is present.


Fig. 16.33
After-image test (Courtesy of Wilmer Eye Institute)


Fig. 16.35
Synoptophore

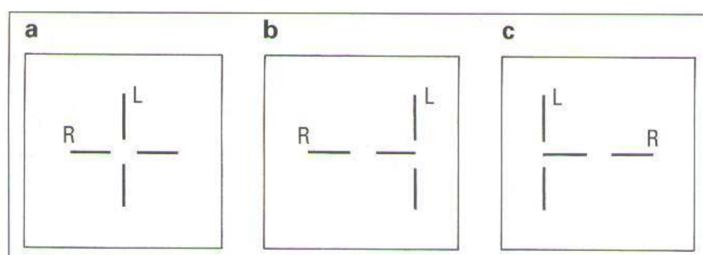


Fig. 16.34
Possible results of the after-image test (*see text*)

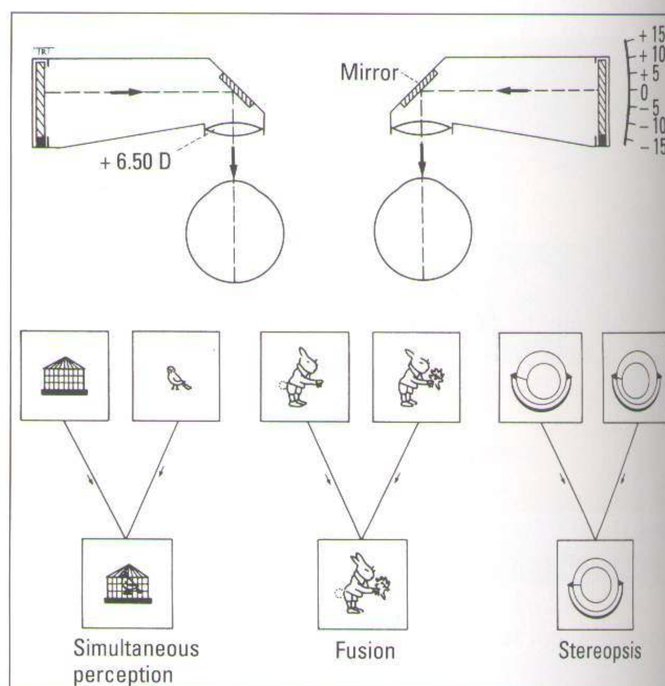


Fig. 16.36
Optical principles of the synoptophore (*see text*)

- In esotropia with ARC, the horizontal after-image (if presented to the right eye) is seen to the left of the vertical image (Fig. 16.34b).
- These findings are reversed in an exotropia (Fig. 16.34c).
- A patient with eccentric fixation will also see a cross. Eccentric fixation is a uniocular condition in which an extrafoveal part of the retina is used for fixation under both binocular and monocular conditions. There is a reorientation of the sensory and motor functions so that this new area usurps the principal visual direction once possessed by the fovea. The fovea of the dominant eye will perceive its after-image as straight ahead in visual space. With regard to the deviated eye, the eccentric area stimulated during the test will also perceive its after-image as straight ahead because that area has 'stolen' the principal visual direction.

Synoptophore

This is an instrument for assessing strabismus and quantifying binocular vision (Fig. 16.35). It can also detect suppression and ARC. The instrument consists of two cylindrical tubes with a mirrored right-angled bend and a +6.50 D lens in each eyepiece (Fig. 16.36, top). This optically sets the testing distance at about 6 metres. Pictures are inserted in a slide carrier situated at the outer end of each tube. The two tubes are supported on columns which enable the pictures to be moved in relation to each other, and any adjustments are indicated on a scale. The synoptophore can measure horizontal, vertical and torsional misalignments.

Grades of binocular vision

Binocular vision is graded on the synoptophore as follows (Fig. 16.36, bottom):

1. **First grade** (simultaneous perception) is tested by introducing two dissimilar but not mutually antagonistic pictures, such as a bird and a cage. The subject is then asked to put the bird into the cage by altering the columns. If the two pictures cannot be seen simultaneously, then either suppression or significant amblyopia is present. The

dissimilar objects are generally not seen in the same position in space. Retinal 'rivalry' will ensure that the image from one eye or the other predominates. However, here one picture is smaller than the other, so that while the small one is seen foveally, the larger one is seen parafoveally (and is thus placed in front of the deviating eye).

2. **Second grade** (fusion) is the ability of the two eyes to produce a composite picture from two similar pictures each of which is incomplete in one small different detail. The classic example is two rabbits, one lacking a tail and the other lacking a bunch of flowers. If fusion is present, one rabbit complete with tail and flowers will be seen. The range of fusion is then tested by moving the arms of the synoptophore so that the eyes have to converge and diverge in order to maintain fusion. It is obvious that the presence of simple fusion without any range is of little value in ordinary life.
3. **Third grade** (stereopsis) is the ability to obtain an impression of depth by the superimposition of two pictures of the same object which have been taken from slightly different angles. The classic example is the bucket which is appreciated in three dimensions.

Detection of ARC

ARC can be detected on the synoptophore as follows:

1. The examiner determines the objective angle of the deviation by presenting each fovea alternately with a target until no movement of the eyes is seen.
2. If the images are seen superimposed with the angle between the arms of the synoptophore equal to the objective angle of squint, then retinal correspondence is normal.
3. If the objective and subjective angles are different, ARC is present. The difference in degrees between the subjective and objective angles is the angle of anomaly. ARC is said to be harmonious when the objective angle equals the angle of anomaly and unharmonious when it exceeds the angle of anomaly. In harmonious ARC the subjective angle is zero (i.e. theoretically there will be no movement on a cover test).

Measurement of deviation

Hirschberg test

This gives a rough objective estimate of the angle of a manifest strabismus in uncooperative patients or when fixation is poor. A pen-torch is shone into the eyes from arm's length and the patient is asked to fixate the light. The corneal reflection of the light will be (more or less) centred in the pupil of the fixating eye, but will be decentred in a squinting eye, in the direction opposite to that of the deviation. The distance of the corneal light reflection from the centre of the pupil is noted. The premise is that every millimetre of deviation is equal to 7° (15 Δ). For example, if the reflex is situated at the temporal border of the pupil (assuming a pupillary diameter of 4 mm), the angle is about 30 Δ (Fig. 16.37); if it is at the limbus, the angle is about 90 Δ (Fig. 16.38). This test is also useful in detecting pseudostrabismus, which may be caused by the following conditions:

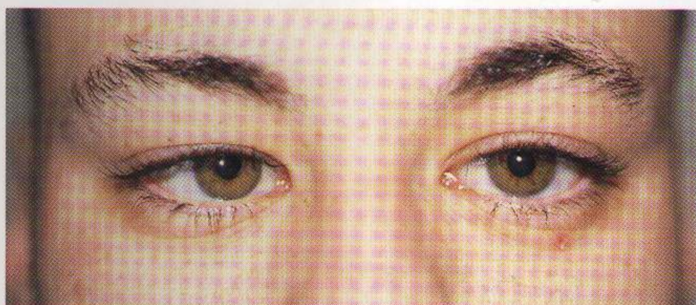


Fig. 16.37
Hirschberg test. The right corneal reflex is near the temporal border of the pupil indicating an angle of about 30 Δ (15°)



Fig. 16.38
Hirschberg test. The left corneal reflex is at the limbus indicating an angle of about 90 Δ (45°)

1. **Pseudo-esotropia**
   a. ***Epicanthic folds*** (*see* Fig. 1.138).
   b. ***Short interpupillary distance*** due to close-set eyes.
   c. ***Negative angle kappa***. Angle kappa is the angle between the visual and anatomical axes (Fig. 16.39b). Normally, the fovea is situated temporal to the posterior pole. The eyes are therefore slightly abducted to achieve bifoveal fixation. A light shone onto the cornea will therefore cause a reflex just nasal to the centre of the cornea in both eyes. This is termed a positive angle kappa; if large it may simulate an exotropia (Fig. 16.39a). A negative angle kappa occurs when the fovea is situated nasal to the posterior pole (high myopia and ectopic fovea). In this situation, the corneal reflex is situated temporal to the centre of the cornea and it may simulate an esotropia.
2. **Pseudo-exotropia**
   a. ***Wide interpupillary distance*** (Fig. 16.40).
   b. ***Positive angle kappa***, as described above.

Krimsky test

In this test, prisms are placed in front of the fixating eye until the corneal light reflexes are symmetrical (Fig. 16.41). It is important to note that a Krimsky test does not dissociate the eyes and only measures a manifest deviation. Since it does not

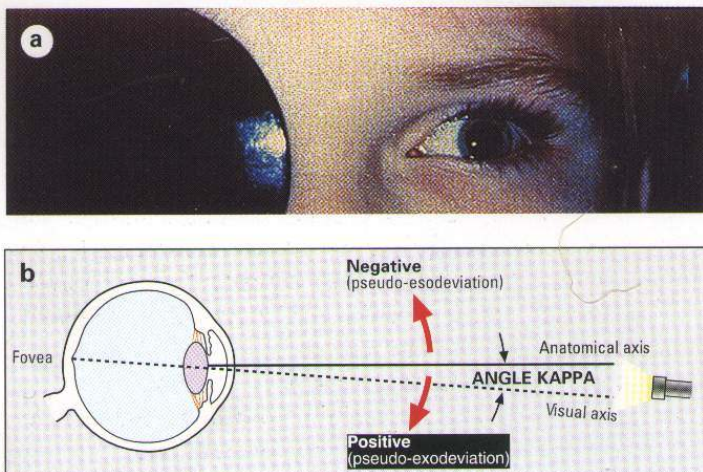


Fig. 16.39
Angle kappa (*see text*) (Courtesy of Wilmer Institute)

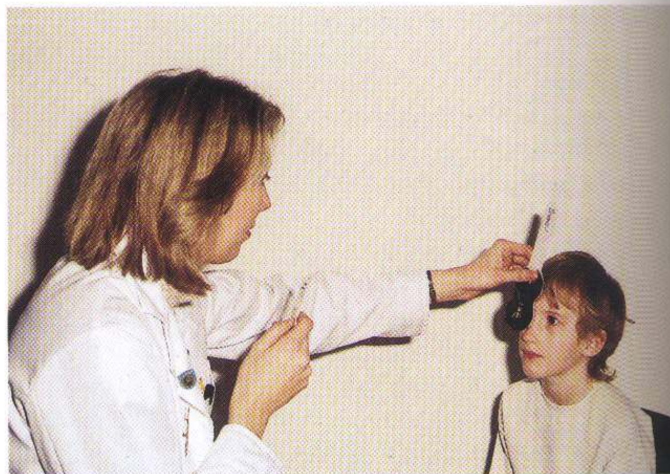


Fig. 16.42
Cover test (*see text*)



Fig. 16.40
Pseudo-exotropia due to a wide interpupillary distance

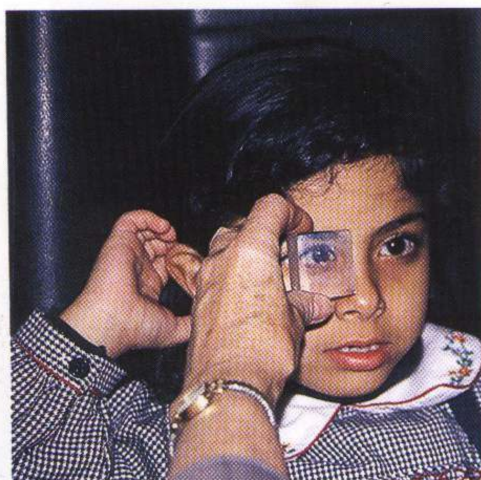


Fig. 16.41
Krimsky test (Courtesy of K. Nischal)

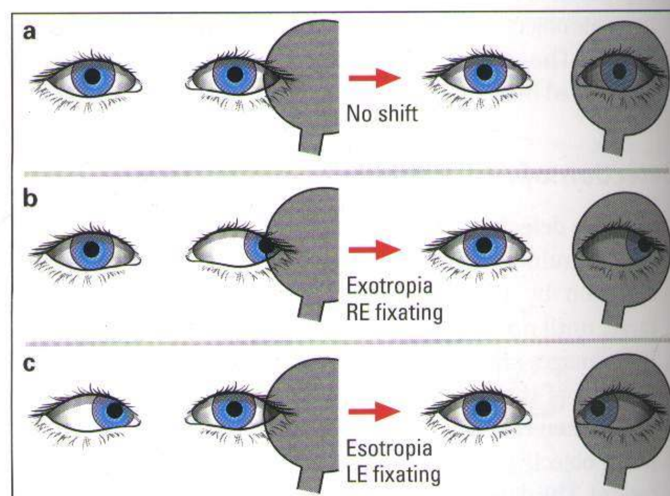


Fig. 16.43
Possible results of the cover test (*see text*)

take into account any latent component, it often underestimates the true size of the deviation.

Cover tests

By far the most accurate assessment of a deviation is with the cover tests. These tests allow the examiner to differentiate tropias from phorias, assess the degree of control of a deviation, and note fixation preference and strength of fixation for each eye. These tests are based on the patient's ability to fixate. Attention and cooperation are also required.

1. Cover–uncover test consists of two parts:

a. Cover test to detect a heterotropia. It should be performed both for near (using an accommodative target) (Fig. 16.42) and for distance as follows:

- The patient fixates a straight-ahead target.
- If a right deviation is suspected, the examiner covers the opposite left eye and notes any movement of the right eye.
- No shift indicates either orthophoria (Fig. 16.43a) or left heterotropia (Fig. 16.43b).
- Adduction of the right eye to take up fixation indicates exotropia and abduction, esotropia (Fig. 16.43c).
- Downward movement indicates hypertropia and

upward movement, hypotropia.
- The test is repeated on the opposite eye.

b. Uncover test to detect a heterophoria. It should be performed both for near (using an accommodative target) and for distance as follows:
- The patient fixates a straight-ahead distant target.
- The examiner covers the right eye and after a few seconds removes the cover (Fig. 16.44).
- No movement indicates orthophoria (Fig. 16.45a), although a keen observer will frequently detect a very slight latent deviation in most normal individuals, as very few people are truly orthophoric.
- If the right eye had deviated while under cover, it will manifest a re-fixation movement on being uncovered.
- Adduction of the right eye indicates exophoria (Fig. 16.45b) and abduction, esophoria (Fig. 16.45c).
- Upward or downward movement indicates a vertical phoria. In the context of latent strabismus, it is never certain whether one eye is 'hypo' or the other eye 'hyper', unlike with a manifest squint.

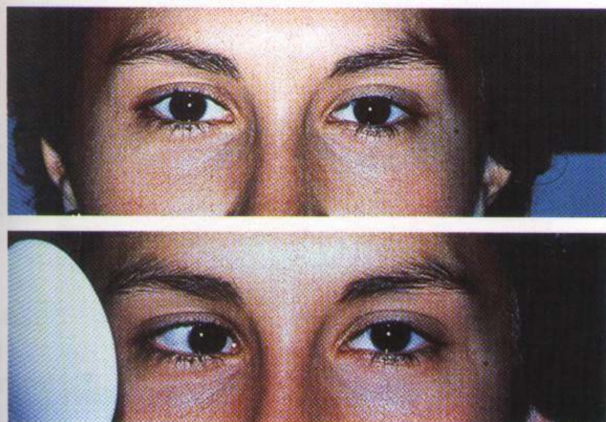


Fig. 16.44
Uncover test (*see text*) (Courtesy of Wilmer Institute)

- The test is repeated for the opposite eye.

> ***NB:*** Most examiners perform the cover test and the uncover test simultaneously, hence the term cover–uncover test.

2. Alternate cover test interrupts binocular fusion mechanisms and reveals the total deviation (phoria plus tropia) (Fig. 16.46). It should be performed after the cover–uncover test because if performed first it will be impossible to differentiate phoria from tropia.
   a. The right eye is covered for about 2 seconds.
   b. The occluder is quickly shifted to the opposite eye for 2 seconds, then back and forth several times.
   c. After the cover is removed, the examiner notes the speed and smoothness of recovery as the eyes return to their pre-dissociated state.
   d. A patient with a heterophoria will have straight eyes before and after the test has been performed (Fig. 16.46a), whereas a patient with a heterotropia will have a manifest deviation (Fig. 16.46b and c).

3. Prism cover test precisely measures the angle of deviation. It is performed as follows:
   a. The alternate cover test is first performed.
   b. Prisms of increasing strength are placed in front of one eye with the base opposite the direction of the deviation (i.e. point the apex of the prism in the direction of the deviation). For example, in a convergent strabismus the prism is held base-out.
   c. The alternate cover test is continuously performed (Fig. 16.47). As stronger prisms are brought in, the amplitude of ocular re-fixation movements gradually decreases.
   d. The end-point is reached when ocular movements are negated; the angle of deviation then equals the strength of the prism.

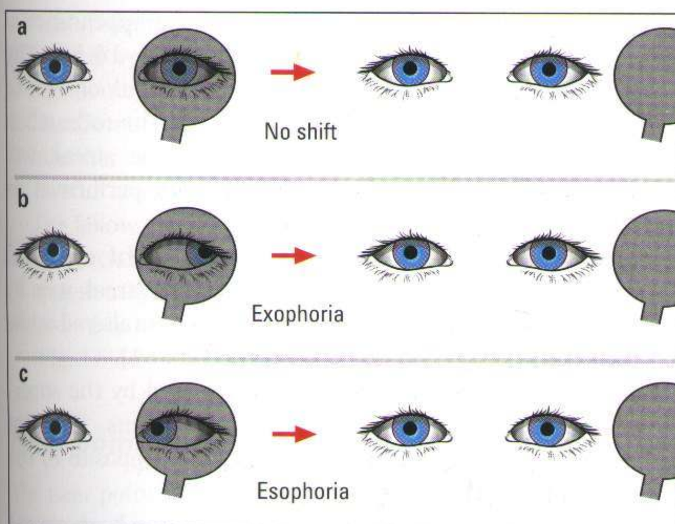


Fig. 16.45
Possible results of the uncover test (*see text*)

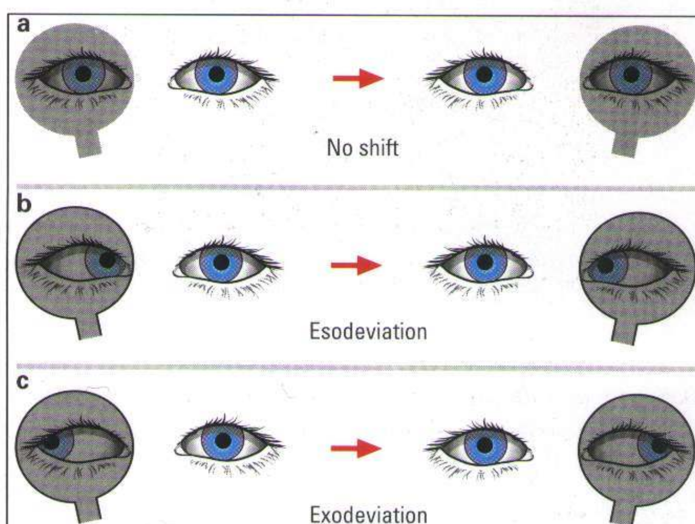


Fig. 16.46
Possible results of the alternate cover test (*see text*)



Fig. 16.47
Prism cover test (*see text*)



Fig. 16.49
Maddox rod test (*see text*) (Courtesy of Wilmer Institute)

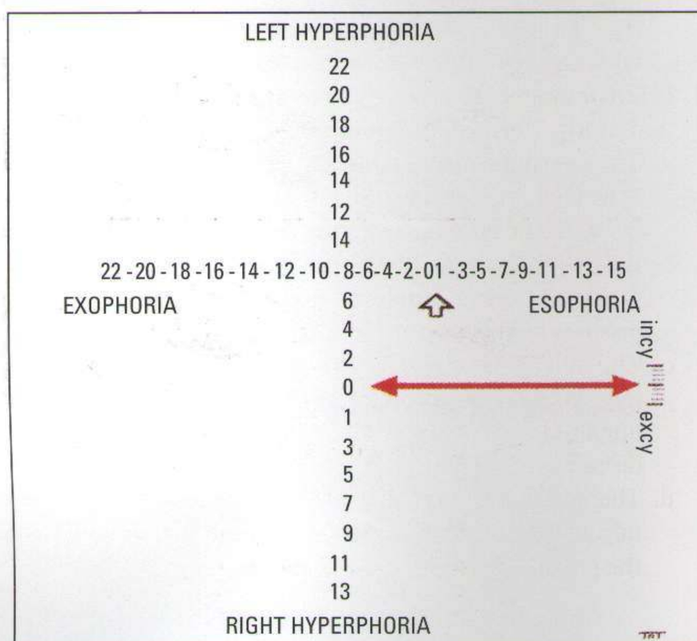


Fig. 16.48
Maddox wing (*see text*)

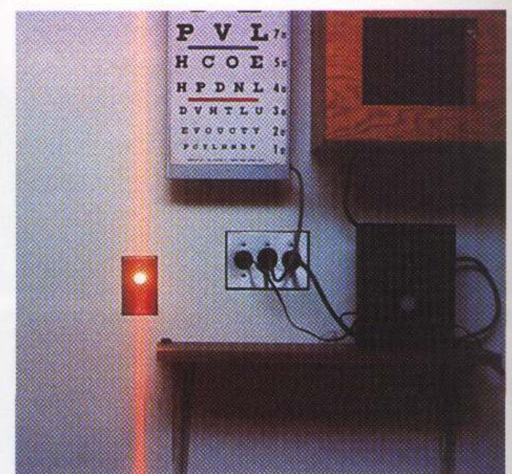


Fig. 16.50
Appearance of a point of light through Maddox rods (Courtesy of Wilmer Institute)

Dissimilar image tests

1. **Maddox wing** dissociates the eyes for near fixation (1/3 m) and measures heterophoria. The instrument is constructed in such a way that the right eye sees only a white vertical arrow and a red horizontal arrow, whereas the left eye sees only horizontal and vertical rows of numbers (Fig. 16.48). Measurements are made as follows:
   - The horizontal deviation is measured by asking the patient to which number the white arrow points.
   - The vertical deviation is measured by asking the patient which number the red arrow intersects.
   - The amount of cyclophoria is determined by asking the patient to move the red arrow so that it is parallel with the horizontal row of numbers.
2. **Maddox rod** consists of a series of fused cylindrical red glass rods (Fig. 16.49) which convert the appearance of a white spot of light into a red streak (Fig. 16.50). The optical properties of the rods cause the streak of light to be at an angle of 90° to the long axis of the rods; when the glass rods are held horizontally, the streak will be vertical and vice versa. The test is performed as follows:
   - The rod is placed in front of the right eye. This dissociates the two eyes because the red streak seen by the right eye cannot be fused with the unaltered white spot of light seen by the left eye (Fig. 16.51).
   - The amount of dissociation is measured by the superimposition of the two images using prisms. The base of the prism is placed in the position opposite to the direction of the deviation.
   - Both vertical and horizontal deviations can be measured in this way but the test cannot differentiate phoria from tropia.

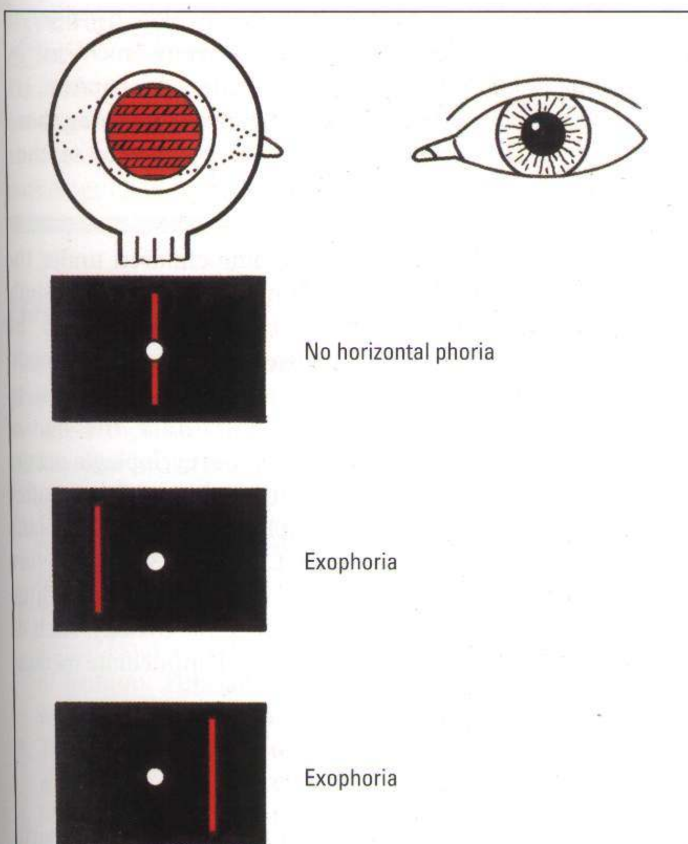


Fig. 16.51
Possible results of the Maddox rod test (*see text*)

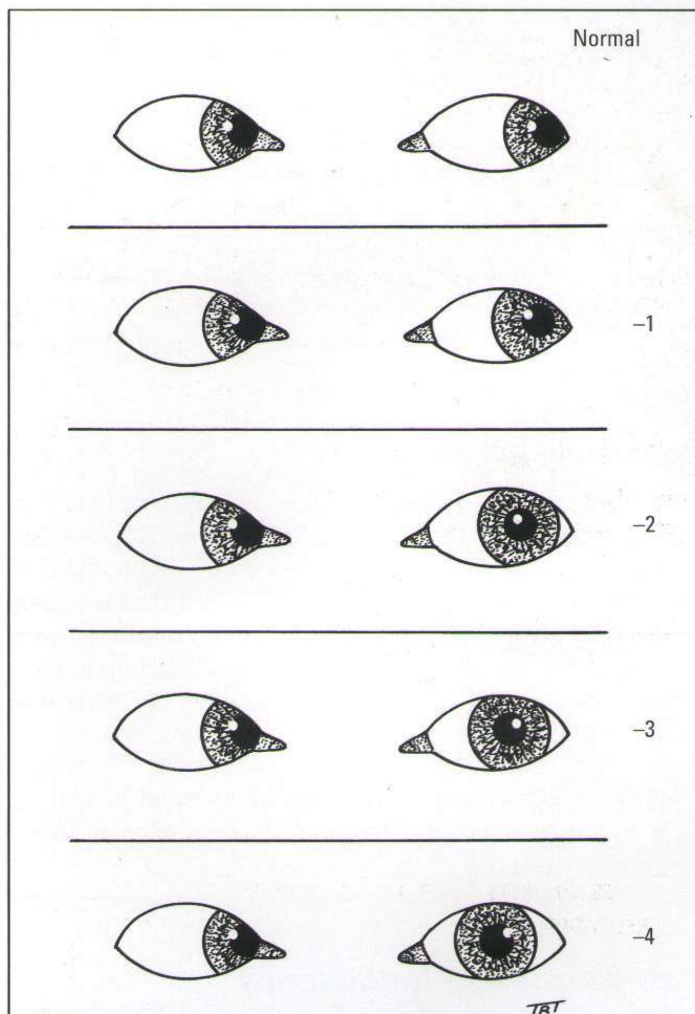


Fig. 16.52
Grading of left lateral rectus underaction



Fig. 16.53
RAF rule (*see text*) (Courtesy of Wilmer Institute)

Motility tests

Ocular movements

Examination of ocular movements involves assessment of smooth pursuit movements followed by that of saccadic movements.

1. **Versions** towards the eight eccentric positions of gaze are tested by asking the patient to follow a target, usually a pen or pen-torch (which offers the advantage of corneal light reflections to aid assessment). They may also be elicited voluntarily, acoustically or by the doll's head manoeuvre.
2. **Ductions** are assessed if reduced ocular motility is noted in either or both eyes. A pen-torch should be used with careful attention to the position of the corneal reflexes. The fellow eye is occluded and the patient asked to follow the torch into various positions of gaze. A simple numeric system may be employed using 0 to denote full movement, and –1 to –4 to denote increasing degrees of underaction (Fig. 16.52).

Near point of convergence

The near point of convergence (NPC) is the nearest point on which the eyes can maintain fixation. It can be measured with the RAF rule, which rests on the patient's cheeks (Fig. 16.53). A target is slowly moved along the rule towards the patient's eyes until one eye loses fixation and drifts laterally (objective NPC). The subjective NPC is the point at which the patient reports diplopia. Normal NPC should be nearer than 10 cm.

Near point of accommodation

The near point of accommodation (NPA) is the nearest point on which the eyes can maintain clear focus. It can also be measured with the RAF rule. The patient fixates a line of print, which is then slowly moved towards the patient until it becomes blurred. The distance at which this is first reported is read off the rule and denotes the NPA. The NPA recedes with the age; when sufficiently far away to render reading difficult without optical correction, this indicates presbyopia. At the age of 20 years the NPA is 8 cm and by the age of 50 years it has receded to 46 cm.

Fusional amplitudes

These measure the efficacy of disjunctive movements. They may be tested with prisms or the synoptophore. Increasingly strong prisms are placed in front of one eye, which will then abduct or adduct (depending on whether the prism is base-in or base-out respectively), in order to maintain bifoveal fixation. When a prism greater than the fusional amplitude is reached, diplopia is reported or one eye drifts the other way. This is the limit of vergence ability.

> ***NB:*** The prism fusion range must be assessed in any patient where there is a risk of diplopia developing after strabismus surgery.

Refraction and fundoscopy

It should be emphasized that dilated fundoscopy is *mandatory* in the context of strabismus, to exclude any underlying ocular pathology such as macular scarring, optic disc hypoplasia or retinoblastoma. Strabismus is often secondary to refractive error. Hypermetropia, astigmatism, anisometropia and myopia may all be associated with strabismus.

Cycloplegia

The commonest refractive error causing strabismus is hypermetropia. Accurate measurement of hypermetropia necessitates effective paralysis of the ciliary muscle (cycloplegia), in order to neutralize the effect of accommodation, which masks the true degree of this refractive error.

1. **Cyclopentolate** affords adequate cycloplegia in most children. The concentration employed is 0.5% under the age of 6 months and 1% thereafter. One drop, repeated after 5 minutes, usually results in maximal cycloplegia within 30 minutes, with recovery of accommodation after 24 hours. The adequacy of cycloplegia can be determined by comparing retinoscopy readings with the patient fixating for distance and then for near. If cycloplegia is adequate, there will be little or no difference. If cycloplegia is incomplete there will be a difference between the two readings and it may be necessary to wait another 15 minutes or to instil another drop.

> ***NB:*** Topical anaesthesia with an agent such as proxymetacaine, prior to instillation of cyclopentalate, is useful in preventing ocular irritation and reflex tearing, thus affording better retention of the cyclopentolate in the conjunctival sac and effective cycloplegia.

2. **Atropine** may be necessary in some children under the age of 4 years with either high hypermetropia or heavily pigmented irides, in whom cyclopentolate may be inadequate. Atropine may be used as drops or ointment, although drops are easier for an untrained person to instil. The concentration is 0.5% under the age of 12 months and 1% thereafter. Maximal cycloplegia occurs at 3 hours, recovery of accommodation starts after about 3 days and is usually complete by 10 days. Atropine is instilled (by the parents) t.i.d. for 3 days before retinoscopy. The parents should be warned to discontinue medication if there are signs of systemic toxicity, such as flushing, fever or restlessness, and seek immediate medical attention.

When to prescribe?

Any significant refractive error should be corrected, particularly in patients with anisohypermetropia or anisoastigmatism associated with amblyopia.

1. **Hypermetropia**. The lowest hypermetropic correction worth prescribing depends on the age and ocular alignment. In the absence of esotropia in a child under the age of 2 years this is +4.00 D, although in an older child it is worth prescribing +2.00 D. However, in the presence of esotropia a correction of +2.00 D should be prescribed, even under the age of 2 years.
2. **Astigmatism**. A cylinder of 1.00 D or more should be prescribed, especially in cases of anisometropia.
3. **Myopia**. The necessity for correction depends on the age of the child. Under the age of 2 years, −5.00 D or more of myopia should be corrected; between the ages of 2 and 4 the amount is −3.00 D. Older children should have correction of even milder degrees of myopia to allow clear distance vision.

Change of refraction

Because refraction changes with age, it is important to check every 6 months. At birth most babies are hypermetropic. After the age of 2 years there may be an increase in hypermetropia and a decrease in astigmatism. Hypermetropia may continue to increase until the age of about 6 years, and then between the ages of 6 and 8 years levels off, subsequently decreasing until the early teenage years. Children under the age of 6 years, with less than +2.50 D, are often virtually emmetropic by the age of 14 years. However, an esotrope under the age of 6 years, with more than +4.00 D, is unlikely to lose sufficient hypermetropia to maintain straight eyes without spectacles.

Investigation of diplopia

The Hess test and the Lees screen plot ocular position as a function of the extraocular muscles and enable differentiation of paretic strabismus caused by neurological pathology from restrictive myopathy such as in thyroid eye disease or a blow-out fracture of the orbit.

Hess test

The screen contains a tangent pattern printed onto a dark-grey background. Red lights that can be individually illuminated indicate the positions of gaze of each of the extraocular muscles.

1. The patient is seated 50 cm from the screen and wears red–green goggles, red lens in front of the right eye, and holds a green 'laser' pointer.
2. The examiner projects a vertical slit of red light onto the screen from a red 'laser' pointer, which is used as the point of fixation. This can now be seen only with the right eye, which therefore becomes the fixating eye.
3. The patient is asked to superimpose their horizontal slit of green light onto the red light.
4. In orthophoria the two lights should be more or less superimposed in all nine positions of gaze.
5. The goggles are then reversed (red filter in front of the left eye) and the procedure is repeated.
6. The relative positions are connected with straight lines.

Lees screen

The apparatus consists of two opalescent glass screens at right-angles to each other, bisected by a two-sided plane mirror which dissociates the two eyes (Fig. 16.54). Each screen has a tangent pattern marked onto the back surface which is revealed only when the screen is illuminated. The test is performed with each eye fixating in turn.



Fig. 16.54
Lees screen (see *text*)

1. The patient faces the non-illuminated screen and fixates the dots in the mirror.
2. The examiner indicates the dot required for the patient to plot.
3. The patient positions the pointer on the non-illuminated screen perceived to be on top of the dot indicated by the examiner.
4. When all of the dots have been plotted the patient is re-positioned to face the other screen and the procedure is repeated.

Interpretation

1. The two charts are compared (Fig. 16.55).
2. The smaller chart indicates the eye with the paretic muscle (right eye).
3. The larger chart indicates the eye with the overacting muscle (left eye).
4. The smaller chart will show its greatest restriction in the main direction of action of the paretic muscle (right lateral rectus).
5. The larger chart will show its greatest expansion in the main direction of action of the yoke muscle (left medial rectus).

Changes with time

Changes with time are also extremely useful as a prognostic guide. For example, in right superior rectus palsy, the Hess chart will show underaction of the affected muscle with an overaction of its yoke muscle (left inferior oblique) (Fig. 16.56a). Because of the great incomitance of the two charts, the diagnosis is straightforward. If the paretic muscle recovers its function, both charts will revert to normal. However, if the paresis persists, the shapes of both charts will change as follows:

- Secondary contracture of the ipsilateral antagonist (right inferior rectus) will show up on the chart as an overaction which will lead to a secondary (inhibitional) palsy of the antagonist of the yoke muscle (left superior oblique), which will show up on the chart as an underaction (Fig. 16.56b). This could lead to the incorrect impression that the left superior oblique was the primary muscle at fault.

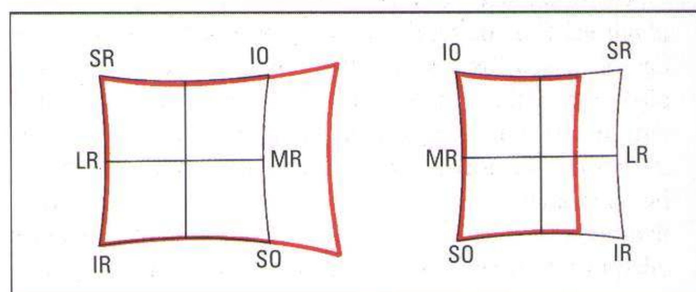


Fig. 16.55
Hess chart of a recent right lateral rectus palsy (see *text*)

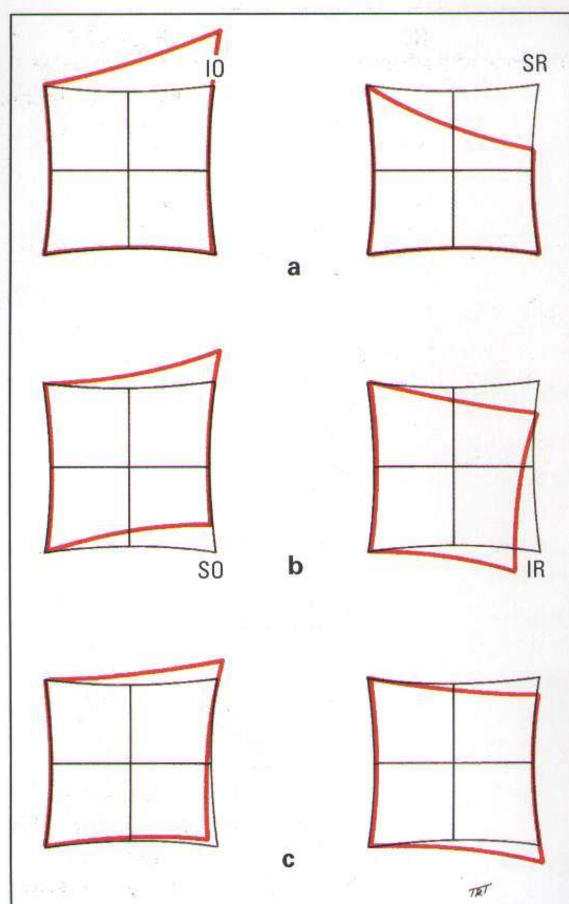


Fig. 16.56
Hess chart showing changes with time of a right superior rectus palsy (*see text*)

- With further passage of time, the two charts become more and more concomitant until it may be impossible to determine which was the primary paretic muscle (Fig. 15.56c).

Clinical examples

It is worth analysing the following examples after gaining a knowledge of ocular motor nerve palsies from Chapter 18.

1. **Left third nerve palsy** shows the following (Fig. 16.57):
   - Contraction of the left chart and expansion of the right.
   - Left exotropia: note that the fixation spots in the inner charts of both eyes are deviated laterally. The deviation is more on the right chart (when the left eye is fixating), indicating that secondary deviation exceeds the primary, as is typical of a paretic squint.
   - Left chart shows underaction of all muscles except the lateral rectus and superior oblique.
   - Right chart shows overactions of all muscles except the medial rectus and inferior rectus, the 'yokes' of the spared muscles.
2. **Right fourth nerve palsy** shows the following (Fig. 16.58):
   - No significant differences in chart size.
   - Right hypertropia: note that the fixation spot of the right inner chart is deviated upwards and the left is deviated downwards. The chart also illustrates that this hypertropia increases on laevoversion, and disappears on dextroversion.
   - Right chart shows underaction of the superior oblique and overaction of the inferior oblique.
   - Left chart shows overaction of the inferior rectus and underaction (inhibitory palsy) of the superior rectus.
3. **Right sixth nerve palsy** shows the following (Fig. 16.59):
   - Contraction of the right chart and expansion of the left.
   - Right esotropia: note that the fixation spot of the right inner chart is deviated nasally.
   - Right chart shows marked underaction of the lateral rectus and slight overaction of the medial rectus.
   - Left chart shows overaction of the medial rectus.

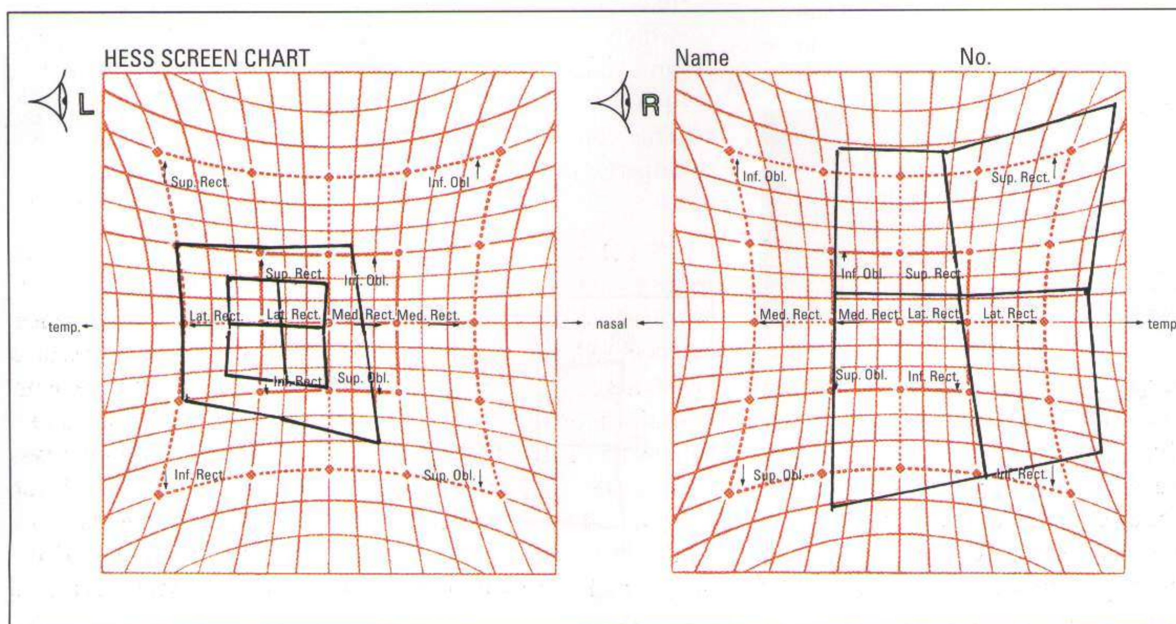
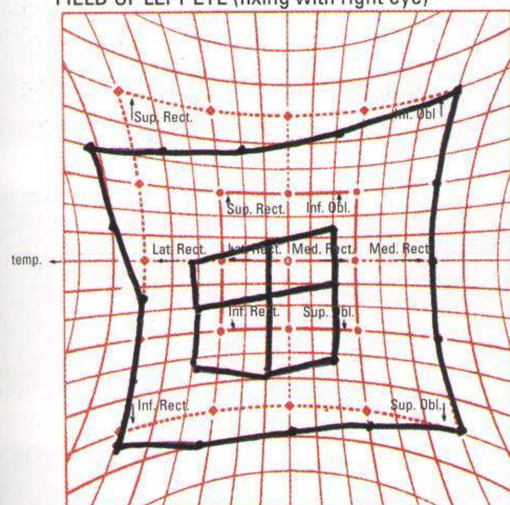


Fig. 16.57
Hess chart of a left third nerve palsy (*see text*)



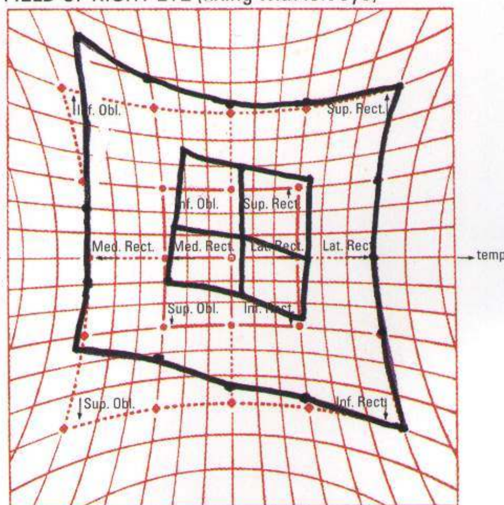
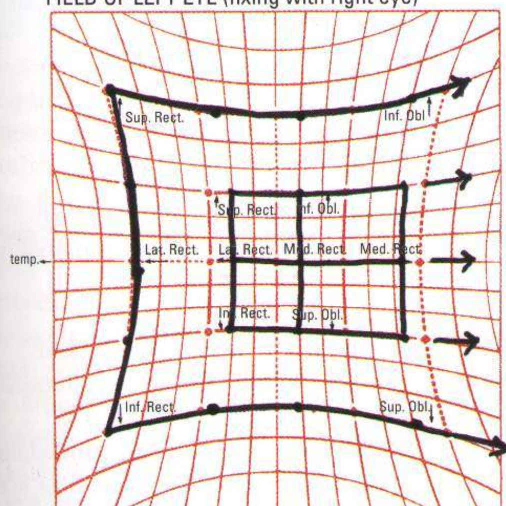


Fig. 16.58
Hess chart of a right fourth nerve palsy (*see text*)



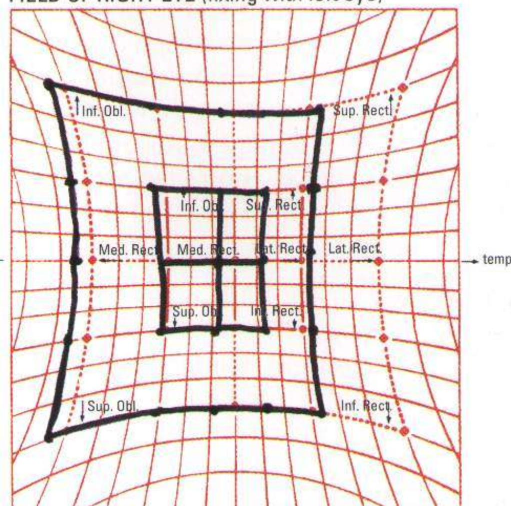


Fig. 16.59
Hess chart of a right sixth nerve palsy (*see text*)

Esotropia

Esotropia (manifest convergent squint) may be comitant or incomitant. In a comitant esotropia the variability of the angle of deviation is within 5 Δ in different horizontal gaze positions. In an incomitant deviation the angle differs in various positions of gaze as a result of abnormal innervation or restriction. Incomitant deviations are discussed later and this section deals only with comitant esotropia. The classification is shown in Table 16.1.

Accommodative esotropia

Near vision involves both accommodation and convergence. Accommodation is the process by which the eye focuses on a near target, by altering the curvature of the crystalline lens. Simultaneously, the eyes converge, in order to fixate bifoveally on the target. Both accommodation and convergence are quantitatively related to the proximity of the target, and have a fairly constant relationship to each other (AC/A ratio) as described previously. Abnormalities of the AC/A ratio are an important cause of certain types of esotropia.

Table 16.1 Classification of esotropia

1. **Accommodative**
   a. Refractive
   - fully accommodative
   - partially accommodative

   b. Non-refractive
   - with convergence excess
   - with accommodation weakness

   c. Mixed accommodative
2. **Non-accommodative**
   - essential infantile
   - microtropia
   - basic
   - convergence excess
   - convergence spasm
   - divergence insufficiency
   - divergence paralysis
   - sensory
   - consecutive
   - acute-onset
   - cyclic

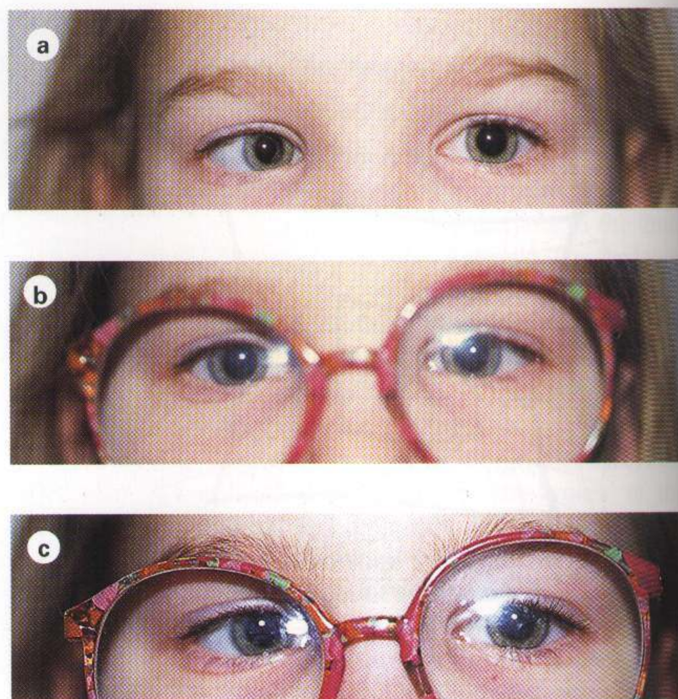


Fig. 16.61
Mixed accommodative esotropia. (**a**) Right esotropia; (**b**) Straight for distance with spectacle correction; (**c**) right esotropia for near with spectacle correction (Courtesy of K. Nischal)

Refractive accommodative esotropia

Here the AC/A ratio is normal and esotropia is a physiological response to excessive hypermetropia, usually between +4.00 and +7.00 D. The considerable degree of accommodation required to focus clearly on even a distant target is accompanied by a proportionate amount of convergence, which is beyond the patient's fusional divergence amplitude. It cannot therefore be controlled, and a manifest convergent squint results. The magnitude of the deviation varies little (usually $<10\ \Delta$) between distance and near. The deviation presents at about the age of 2.5 years (range of 6 months to 7 years).

1. **Fully accommodative** is completely eliminated by optical correction of hypermetropia (Fig. 16.60).
2. **Partially accommodative** is reduced, but not eliminated by correction of hypermetropia.

Non-refractive accommodative esotropia

This is associated with a high AC/A ratio in which a unit increase of accommodation is accompanied by a disproportionately large increase of convergence in the absence of significant hypermetropia. The two types are:

1. **Convergence excess**, which is characterized by:
   - High AC/A ratio due to increased AC (accommodation is normal, convergence is exaggerated).
   - Normal near point of accommodation.
   - Straight eyes for distance, but esotropia for near.
2. **With defective accommodation** (hypoaccommodative), which is characterized by:
   - High AC/A ratio due to decreased A (accommodation is weak, necessitating increased effort, which is accompanied by a strong convergence response).
   - Remote near point of accommodation.
   - Extra accommodative effort is required for near, resulting in convergence excess.

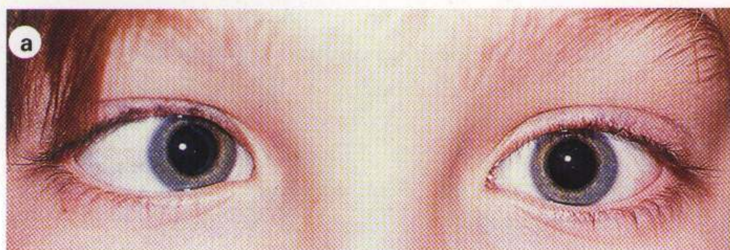




Fig.16.60
Refractive fully accommodative esotropia. (**a**) Right esotropia; (**b**) straight eyes with spectacle correction (Courtesy of K. Nischal)

Mixed accommodative esotropia

Hypermetropia and a high AC/A ratio may coexist, resulting in esotropia for distance, which increases markedly (>10 Δ) on near fixation. The distance deviation is usually corrected by spectacles (Fig. 16.61a and b), but the patient will still have an esotropia for near (Fig. 16.61c), unless wearing bifocals.

Management

1. **Refractive error** should be corrected, as previously described. In children under the age of 6 years, the full cycloplegic refraction revealed on retinoscopy should be prescribed (with a deduction only for the working distance). In the fully accommodative refractive esotrope this will control the deviation for both near and distance. After the age of 8 years, retinoscopy should be performed without cycloplegia and the maximal amount of 'plus' that can be tolerated (the manifest hypermetropia) prescribed.
2. **Bifocals** may be prescribed if there is accommodative esotropia for near (high AC/A ratio). Bifocals relieve accommodation (and thereby accommodative convergence), thus allowing the child to maintain bi-foveal fixation/ocular alignment at near. The minimum 'plus' required to achieve this is used. The most satisfactory form of bifocals is the executive type in which the intersection crosses the lower border of the pupil. The strength of the lower segment should be gradually reduced and eliminated by the early teenage years. The ultimate prognosis for complete withdrawal of spectacles is related to the magnitude of the AC/A ratio and also the degree of hypermetropia and associated astigmatism. Spectacles may be needed only for close work.
3. **Miotic therapy** can be used short-term in children with accommodative esotropia due to a high AC/A ratio, who will not wear spectacles. The initial dose is 0.125% ecothiopate iodide once daily or 4% pilocarpine q.i.d. for 6 weeks. If this is effective, the strength and frequency can be gradually reduced to a minimal effective dose. The formation of iris cysts induced by ecothiopate can be prevented by the simultaneous administration of 2.5% phenylephrine drops twice daily. Miotic therapy works by inducing 'peripheral' accommodation (i.e. direct stimulation of the ciliary muscle rather than that mediated by the third cranial nerve). Less accommodative effort is therefore required by the patient for near vision and thereby less accommodative convergence is induced. It may, however, result in blurred distance vision.
4. **Treatment of amblyopia**, as previously described, is very important before contemplating surgery.
5. **Surgery** may be considered if spectacles do not fully correct the deviation, after every attempt has been made to treat amblyopia. The principle of surgery involves weakening of the medial recti, the muscles active in ocular convergence.
   - Bilateral medial rectus recessions are performed in patients with equal vision in both eyes, in which the deviation for near is greater than that for distance.
   - If there is no significant difference between distance and near measurements, and equal vision in both eyes, some perform medial rectus recession combined with lateral rectus resection, whereas others prefer bilateral medial rectus recessions.
   - Recession–resections on the amblyopic eye are performed in patients with residual amblyopia.

Essential infantile esotropia

Essential infantile esotropia is an idiopathic esotropia developing within the first 6 months of life in an otherwise normal infant with no significant refractive error and no limitation of ocular movements.

Signs

- The angle is usually fairly large (>30 Δ) and stable (Fig. 16.62).
- Fixation in most infants is alternating in the primary position and cross-fixating in side gaze, so that the child uses the right eye in left gaze (Fig. 16.63b) and the left eye in right gaze (Fig. 16.63a). Such cross-fixation may give a false impression of bilateral abduction deficits, such as with bilateral sixth nerve palsy. Abduction can, however, usually be demonstrated either by the doll's head manoeuvre or rotating the child. Should these fail, uniocular patching for a few hours will often unmask the ability of the other eye to abduct (Fig. 16.63c).
- Nystagmus, if manifest, is usually horizontal (it may be latent, or manifest-latent).
- The refractive error is usually normal for the age of the child (about +1.50 D).



Fig. 16.62
Essential infantile esotropia

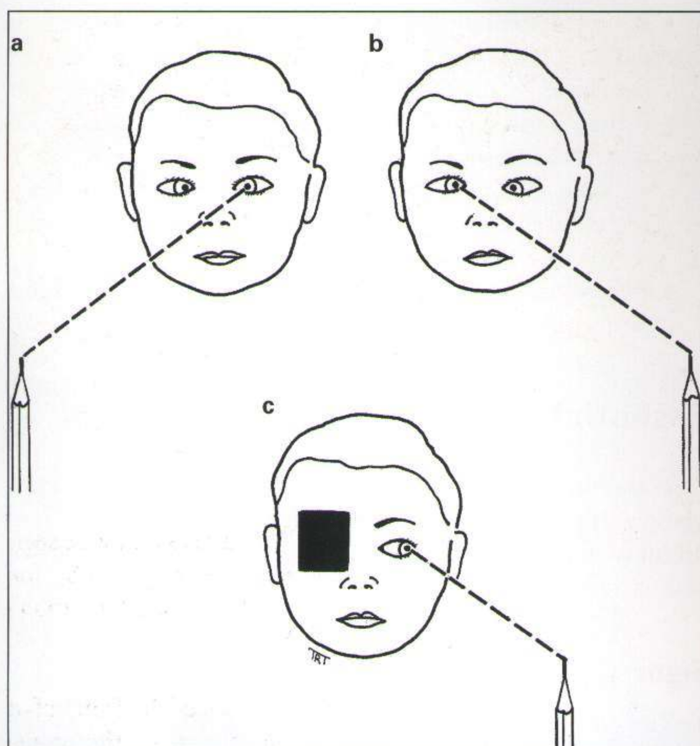


Fig. 16.63
Cross-fixation in essential infantile esotropia (Courtesy of Wilmer Institute)

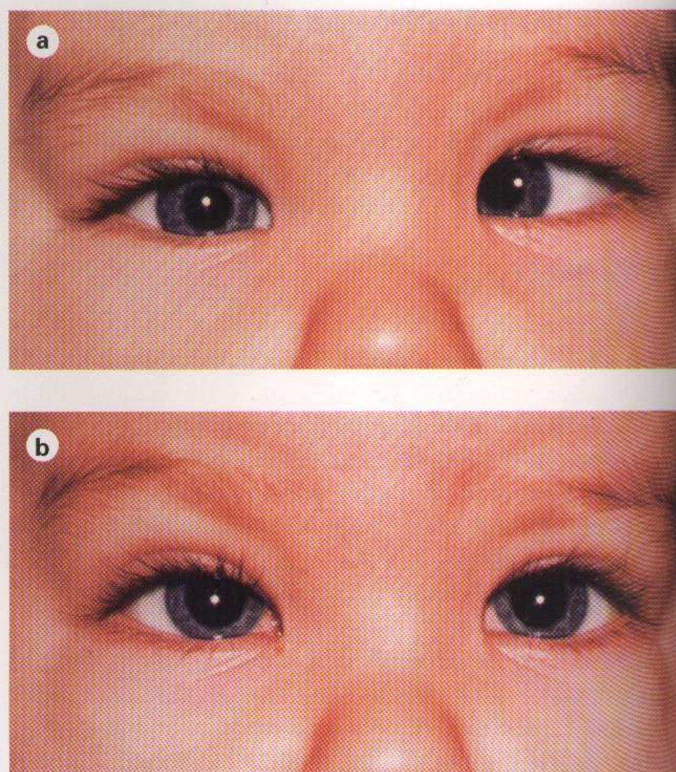


Fig. 16.64
(a) Essential infantile esotropia; **(b)** after appropriate surgery

- Asymmetry of optokinetic nystagmus.
- Inferior oblique overactions may be present initially or develop later.
- Dissociated vertical deviation develops in 80% by the age of 3 years (*see* below).
- Poor potential for BSV.

Differential diagnosis

- Congenital bilateral sixth nerve palsy, which can be excluded as described above.
- Sensory esotropia due to organic eye disease.
- Nystagmus blockage syndrome in which convergence dampens a horizontal nystagmus.
- Duane syndrome types I and III.
- Möbius syndrome.
- Strabismus fixus.

Initial management

Ideally, the eyes should be surgically aligned by the age of 12 months, and at the very latest by the age of 2 years, but only after amblyopia or significant refractive errors have been corrected. The initial procedure is recession of both medial recti. Very large angles may require recessions of 6.5 mm or more. Any associated overactions of the inferior obliques should also be addressed. An acceptable goal is alignment of the eyes to within 10 Δ esotropia, associated with peripheral fusion (combats diplopia) and central suppression (combats confusion) (Fig. 16.64). This small-angle residual strabismus is often stable, even though bifoveal fusion is not achieved.

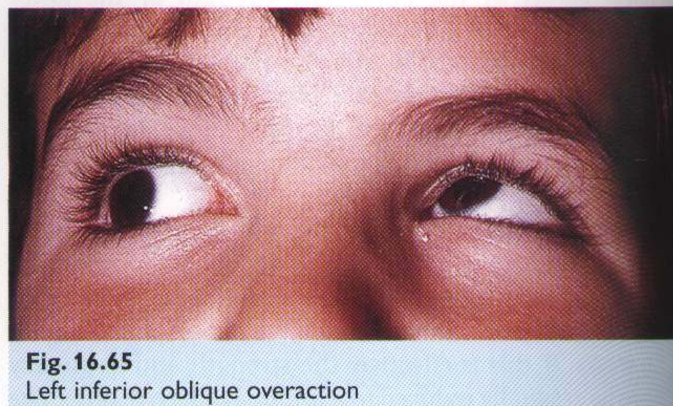


Fig. 16.65
Left inferior oblique overaction

Subsequent management

1. **Undercorrection** may require further recession of the medial recti or resection of one or both lateral recti.
2. **Inferior oblique overaction** may develop subsequently, most commonly at age 2 years (Fig. 16.65). The parents should therefore be warned that further surgery may be necessary despite an initially good result. Initially unilateral, it frequently becomes bilateral within 6 months. Inferior oblique weakening procedures include myotomy, myectomy and recession (*see* later).
3. **Dissociated vertical deviation** (DVD) may appear several years after the initial surgery, particularly in children with nystagmus. It is characterized by the following:

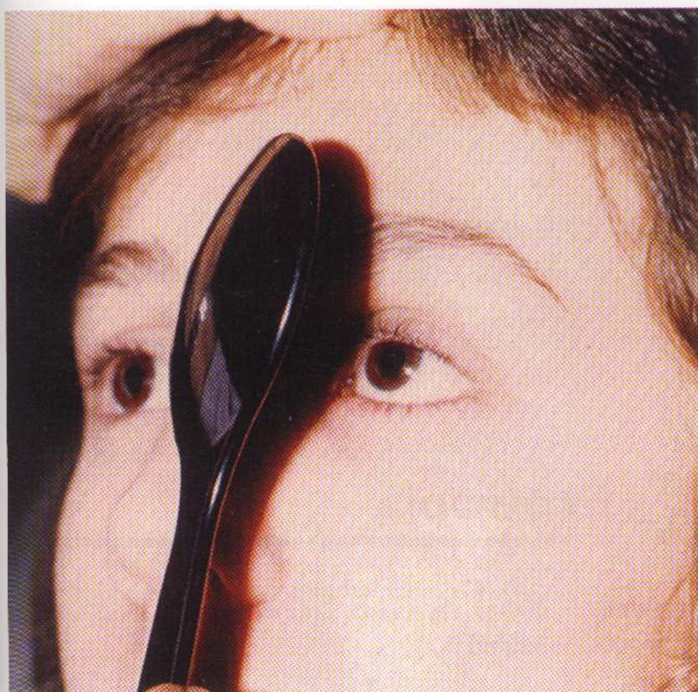

Fig. 16.66
Dissociated vertical deviation (*see text*)


Fig. 16.67
Right microtropia

- Up-drift with excyclodeviation of the eye when under cover or during periods of visual inattention (Fig. 16.66).
- When the cover is removed the affected eye will move down without a corresponding down-drift of the other eye. Thus DVD does not obey Hering law. Although it is usually bilateral, it may be asymmetrical. Surgical treatment is indicated when the condition is cosmetically unacceptable. Superior rectus recession with or without a Faden procedure (*see* later) and/or inferior oblique anterior transposition are popular operations for DVD, although full correction is seldom possible.

4. **Amblyopia** subsequently develops in about 50% of cases.
5. **An accommodative element** should be suspected if the eyes are initially straight, or almost straight after surgery and then start to reconverge. It is therefore important to perform repeated refractions on all children and to correct any new accommodative elements accordingly.

Microtropia

Microtropia (monofixation syndrome) may be primary or follow surgery for a large deviation.

Signs

1. **Anisometropia** in nearly all cases, commonly with hypermetropia or hypermetropic astigmatism.
2. **Very small angle** measuring 8 Δ or less, which may or may not be detectable on cover testing (Fig. 16.67).
3. **Central suppression scotoma** of the deviating eye that prevents confusion may be detected by the following:
   a. ***Bagolini striated glasses*** in which a cross may be seen but there is a gap in the oblique line perceived by the microtropic (scotomatous) eye, at the point of intersection (*see* Fig. 16.32d).
   b. ***4Δ base-out test***
      - When a 4Δ base-out prism is placed in front of a normal eye, the sudden displacement of the image from the fovea to a parafoveal temporal point on the retina will elicit a refixation movement.
      - No movement will be seen in a microtropic eye, since the image is shifted within the central suppression scotoma.
      - In accordance with Hering law, the fellow eye moves outward when the eye under the prism refixates and subsequently performs a slow fusional movement in the opposite direction to correct for its image displacement.
      - If a central scotoma has impaired foveal function, neither movement occurs.
4. **Other features** include ARC, normal or almost normal peripheral fusional amplitudes and reduced stereopsis.

Treatment

This consists of spectacle correction of anisometropia and occlusion for amblyopia but is almost never successful in restoring bifoveal fixation.

Other non-accommodative esotropias

Basic

1. **Signs**
   - No significant refractive error.
   - Deviation is equal for near and distance.
2. **Treatment** is surgical.

Convergence excess

1. **Signs**
   - No significant refractive error.
   - Orthophoria or small esophoria for distance.

- *Esotropia for near but normal or low AC/A ratio.*
- Normal near point of accommodation.

2. **Treatment** involves bilateral medial rectus recessions.

Cyclic oculomotor spasm

This is an intermittent phenomenon which is usually hysterical but may occasionally have an organic cause such as trauma or a posterior fossa tumour.

1. **Signs** during an attack are:
   - Esotropia due to sustained convergence.
   - Pseudo-myopia due to accommodative spasm.
   - Bilateral miosis.
2. **Treatment** is with cycloplegics and bifocals.

Divergence insufficiency

This typically affects healthy young adults.

1. **Signs**
   - Intermittent or constant esotropia for distance.
   - Minimal or no deviation for near.
   - Full abduction bilaterally.
   - Fusional divergence amplitudes may be reduced.
   - Absence of neurological disease.
2. **Treatment** is with prisms until spontaneous resolution or bilateral lateral rectus resections in persistent cases.

Divergence paralysis

This may present at any age and may be difficult to differentiate from unilateral or bilateral sixth nerve palsy, but is usually comitant. It is characterized by:

- Esotropia which is unchanged or may decrease on lateroversion, unlike a sixth nerve palsy.
- Fusional divergence amplitudes are either severely reduced or absent.
- Underlying neurological disease is present such as head trauma, intracranial space-occupying lesions and cerebrovascular accidents.

Sensory esotropia

This is caused by a unilateral reduction in visual acuity which interferes with or abolishes fusion, such as cataract, optic atrophy or hypoplasia, toxoplasma retinochoroiditis or retinoblastoma.

> ***NB:*** Fundus examination under mydriasis is therefore essential in all children with strabismus.

Consecutive esotropia

This follows surgical overcorrection of an exodeviation. Unless the deviation is very large, surgery should be postponed for several months because spontaneous improvement may occur.

Acute-onset esotropia

This is usually due to a sudden decompensation of esophoria or microtropia. The patient complains of diplopia and it is important to exclude sixth nerve palsy or divergence paralysis.

Cyclic esotropia

This is a very rare condition characterized by alternating manifest esotropia and orthophoria, each lasting 24 hours. The condition may persist for months or years and the patient may eventually develop a constant esotropia requiring surgery.

Exotropia

Exotropia (manifest divergent squint) may be constant or intermittent (Table 16.2)

Constant exotropia

Congenital exotropia

1. **Presentation** is at birth, in contrast to infantile esotropia.
2. **Signs**
   - Normal refraction.
   - Large and constant angle.
   - DVD may be present.
3. **Neurological anomalies** are frequently present, in contrast to infantile esotropia.
4. **Treatment** is mainly surgical and consists of bilateral lateral rectus recessions, usually combined with resection of one or both medial recti, depending on the angle.

Other types

1. **Sensory** exotropia is the result of monocular or binocular visual impairment by acquired lesions, such as cataract or other opacities of the media, in children over the age of 5 years or in adults (Fig. 16.68). Treatment consists of correction of the visual deficit, if possible, followed by surgery, if necessary.

Table 16.2 Classification of exotropia

1. **Constant**
   - congenital
   - sensory
   - consecutive
2. **Intermittent**
   - basic
   - convergence weakness
   - divergence excess



Fig. 16.68
Left sensory exotropia due to a dense cataract

2. **Consecutive** exotropia follows surgical correction of an esodeviation.

Intermittent exotropia

1. **Presentation** is most frequently at around 2 years with exophoria, which breaks down to exotropia under conditions of visual inattention, bright light (resulting in reflex closure of the affected eye), fatigue or ill health. With the passage of time, the deviation tends to become less well controlled.
2. **Clinical types**
   a. ***Basic*** exotropia where the angle of deviation is the same for distance and near fixation (Fig. 16.69).
   b. ***Convergence weakness*** tends to occur in older children and adults. The angle of deviation is greater for near fixation (Fig. 16.70). It may be associated with acquired myopia.
   c. ***Divergence excess***, in which the angle of deviation is greater for distance (Fig. 16.71). This type of exotropia may be true or simulated.

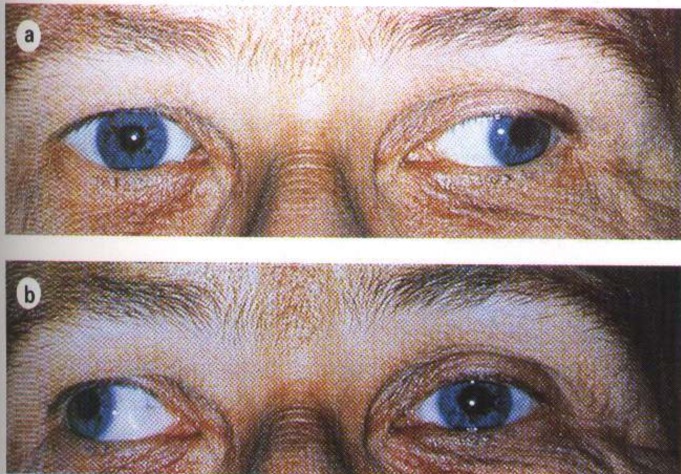
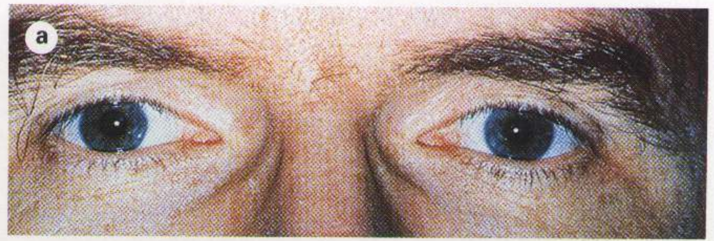


Fig. 16.69
Basic exotropia. Equal deviation for distance (**a**) and near (**b**) (Courtesy of Wilmer Institute)



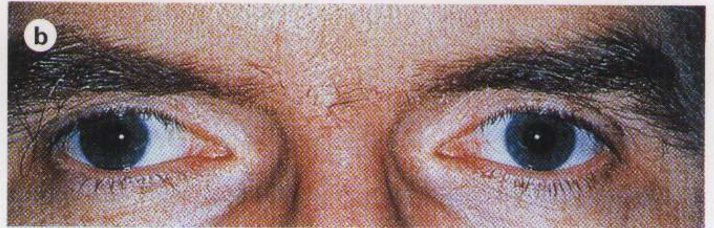


Fig. 16.70
Convergence weakness exotropia. Greater deviation for near (**b**) than distance (**a**) (Courtesy of Wilmer Institute)





Fig. 16.71
Divergence excess exotropia. Greater deviation for distance (**a**) than near (**b**) (Courtesy of Wilmer Eye Institute)

- True type in which the angle for near is consistently less than that for distance.
- Simulated type is associated with a high AC/A ratio. The deviations for near and distance are similar when the near angle is remeasured with the patient looking through a +3.00 D lens or after a period of uniocular occlusion.

Management

1. **Spectacle correction** in myopic patients may, in some cases, control the deviation by stimulating accommodation and, with it, convergence.
2. **Orthoptic treatment** consisting of occlusion therapy, diplopia awareness and improvement of fusional convergence may also be useful in selected cases.
3. **Surgery** is necessary in most patients by about the age of 5 years. Some advocate recessions of both lateral recti,

whereas others recommend bilateral surgery only for those patients with divergence excess, preferring recession–resection where the distance–near measurements are the same.

Special syndromes

Duane syndrome

The hallmark of Duane syndrome is retraction of the globe on attempted adduction caused by co-contraction of the medial and lateral recti. The condition is often bilateral, although frequently involvement of one eye may be so subtle as to go unnoticed. Some children have associated congenital defects; the most common is perceptive deafness with associated speech disorder.

Classification (Huber)

1. **Type I**, the most common, is characterized by:
   - Limited or absent abduction.
   - Normal or mildly limited adduction.
   - In the primary position, straight or slight esotropia.
2. **Type II**, the least common, is characterized by:
   - Limited adduction.
   - Normal or mildly limited abduction.
   - In primary position, straight or slight exotropia.
3. **Type III**, is characterized by:
   - Limited adduction and abduction.
   - In the primary position, straight or slight esotropia.

Other features

Features that may occur in each of the three subgroups include the following:

- On attempted adduction there is retraction of the globe, produced by co-contraction of the medial and lateral recti, with resultant narrowing of the palpebral fissure (Fig. 16.72a). The degree of globe retraction may vary from gross to almost imperceptible. On attempted abduction, the palpebral fissure opens and the globe assumes its normal position (Fig. 16.72b).
- An up-shoot or down-shoot in adduction is seen in some patients. It has been suggested that this is a 'bridle' or 'leash' *phenomenon, produced by a tight lateral rectus muscle* which slips over or under the globe and produces an *anomalous vertical movement of the eye. However, recent* studies with MRI have shown that this is not always the case.

Management

In most cases the eyes are straight in the primary position and there is no amblyopia. Surgery is indicated if the eyes are not straight in the primary position and the patient has to adopt an abnormal head posture to achieve fusion. Surgery may also be necessary for cosmetically unacceptable up-shoots, down-shoots or severe globe retraction. Amblyopia, when present, is usually the result of anisometropia rather than strabismus.

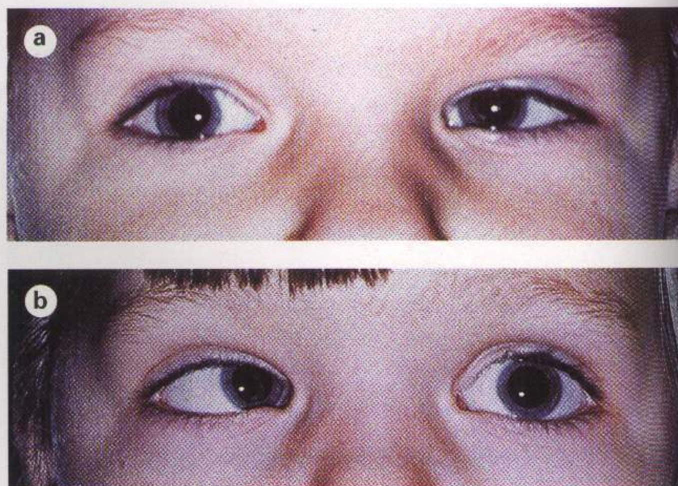


Fig. 16.72
Left Duane retraction syndrome (*see text*) (Courtesy of Wilmer Institute)

> ***NB:*** The lateral rectus of the involved side should not be resected, as this increases retraction.

Brown syndrome

Brown syndrome is usually congenital but occasionally may be acquired:

Classification

1. **Congenital**
   - Idiopathic.
   - Congenital click syndrome where there is impaired movement of the superior oblique tendon through the trochlea.
2. **Acquired**
   - Iatrogenic damage to the trochlea or superior oblique tendon.
   - Inflammation of the tendon which may be caused by rheumatoid arthritis, pansinusitis and scleritis.

Clinical features

A right Brown syndrome has the following characteristics:

1. ***Major signs***
   - Usually straight in the primary position (Fig. 16.73a).
   - Limited right elevation in adduction and occasionally also in the midline (Fig. 16.73b).
   - Usually normal right elevation in abduction (Fig. 16.73c).
   - Minimal or no superior oblique overaction.
   - Positive forced duction test on elevating the globe in adduction.







Fig. 16.73
Right Brown syndrome (*see text*)

2. Variable signs

- Down-shoot in adduction.
- Hypotropia in primary position.
- Anomalous head position with ipsilateral tilt and chin elevation.

Differential diagnosis

1. **Inferior oblique palsy** has more vertical deviation in the primary position, superior oblique overaction, 'A' pattern and negative forced duction test.
2. **Monocular elevation deficit** is characterized by inability to elevate one eye in any position (Fig. 16.74).

Management

1. **Congenital** cases do not usually require treatment. Indications for surgery include primary position hypotropia and/or an anomalous head posture. The recommended procedure for congenital cases is superior oblique weakening.







Fig. 16.74
Right monoelevation deficit showing defective elevation in all positions

2. **Acquired** cases may benefit from steroids, either orally or by injection near the trochlea, together with treatment of the underlying cause.

Möbius syndrome

Möbius syndrome is a very rare congenital, sporadic condition.

1. **Ocular features**
   - Horizontal gaze palsy is present in 50% of cases but vertical gaze is usually intact.
   - Bilateral sixth nerve palsies (Fig. 16.75a).
   - Esotropia in the primary position in 50% of cases; the remainder are straight.
   - A pseudo-esotropia may be seen in children who learn to cross-fixate because adduction and convergence are intact.
2. **Systemic features**
   - Bilateral facial palsy, which is usually asymmetrical and often incomplete, giving rise to a mask-like facial expression and problems with lid closure (Fig. 16.75b).
   - Paresis of the ninth and twelfth cranial nerves. The latter results in atrophy of the tongue (Fig. 16.75c).
   - Mild mental handicap.
   - Limb anomalies.

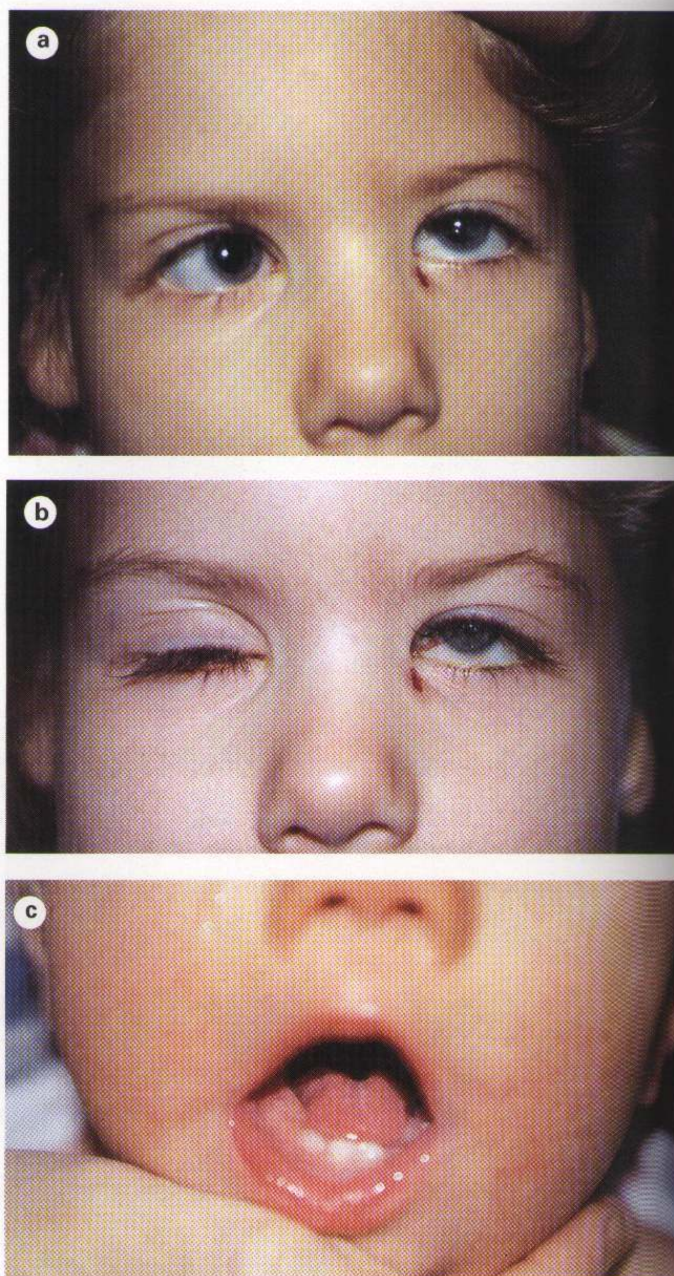


Fig. 16.75
Möbius syndrome. (**a**) Defective left abduction due to a sixth nerve palsy; (**b**) facial palsy; (**c**) atrophic tongue (Courtesy of K. Nischal)

Fibrosis syndromes

1. **Strabismus fixus** is a very rare condition, in which both eyes are fixed, by fibrous tightening of the medial recti (convergent strabismus fixus), or the lateral recti (divergent strabismus fixus).
2. **Congenital fibrosis of extraocular muscles syndrome** is a rare, autosomal-dominant, non-progressive disorder characterized by hypoplasia and fibrosis of extraocular muscles. It is characterized by bilateral congenital ptosis and restrictive external ophthalmoplegia. In the 'primary' position each eye is fixed below the horizontal, by about 10°. The hypotropic eye may be secondarily exotropic (Fig. 16.76), esotropic or neutral.


Fig. 16.76
Exotropia in congenital fibrosis of extraocular muscle syndrome (Courtesy of Wilmer Institute)

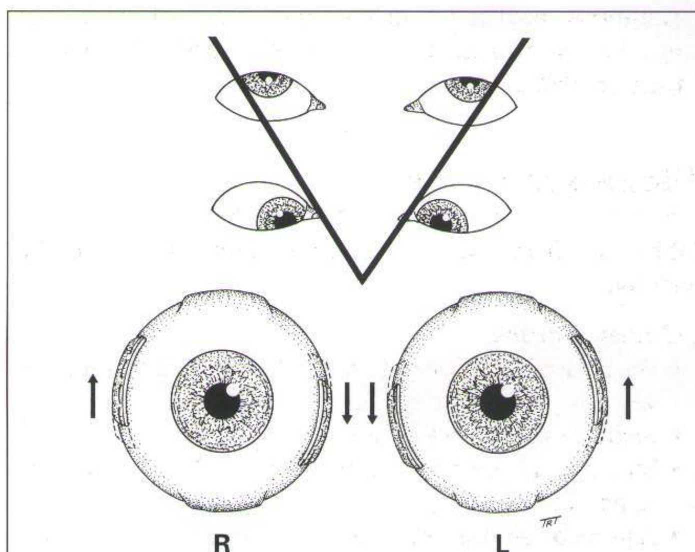


Fig. 16.77
Treatment of 'V' pattern by downward transposition of the medial recti or upward transposition of the lateral recti (see *text*)

The degree of residual horizontal movement varies from full to completely restricted. However, vertical movements are always severely restricted with inability to elevate the eyes above the horizontal plane.

Alphabet patterns

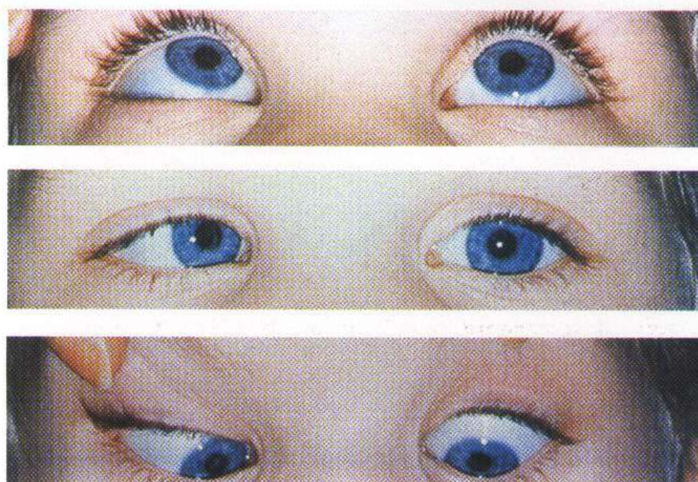
Horizontal deviations can vary when measured in the primary position, upgaze and downgaze, regardless of whether a deviation is comitant or incomitant.

'V' pattern

'V' pattern is significant when difference between upgaze and downgaze is >15 Δ.

Causes

- Brown syndrome.
- Inferior oblique overaction associated with fourth nerve palsy.
- Superior oblique underaction with subsequent inferior oblique overaction which is seen in infantile esotropia as well as other childhood esotropias. The eyes are often straight in upgaze, with a marked esodeviation in downgaze.
- Superior rectus underaction.
- Lateral rectus overaction.
- Craniofacial anomalies which are associated with shallow orbits and downslanting palpebral fissures. There is often a gross exotropia in upgaze and the eyes are almost straight in downgaze.


Fig. 16.78
'V' pattern esotropia (Courtesy of Wilmer Institute)

Treatment

Without oblique muscle dysfunction (Fig. 16.77):

1. **'V' pattern esotropia** (Fig. 16.78) is treated by bilateral medial rectus recessions and downward transposition of the tendons.
2. **'V' pattern exotropia** (Fig. 16.79) is treated by bilateral lateral rectus recessions and upward transposition.

'A' pattern

'A' pattern is significant if the difference between upgaze and downgaze is >10 Δ.



Fig. 16.79
'V' pattern exotropia (Courtesy of Wilmer Institute)

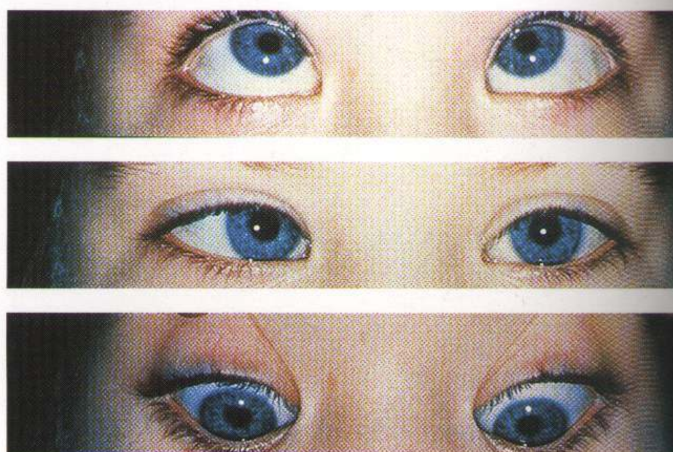


Fig. 16.81
'A' pattern esotropia (Courtesy of Wilmer Institute)

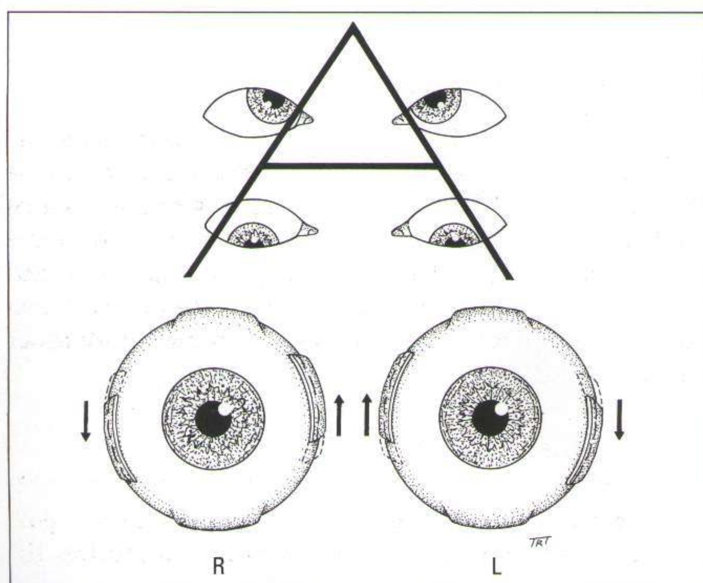


Fig. 16.80
Treatment of 'A' pattern by upward transposition of medial recti or downward transposition of lateral recti (*see text*)

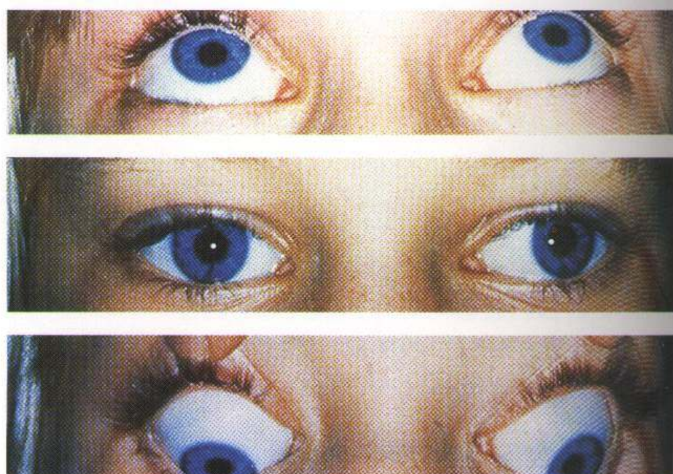


Fig. 16.82
'A' pattern exotropia (Courtesy of Wilmer Institute)

Causes

- Primary superior oblique overaction which is usually associated with exodeviation in the primary position of gaze.
- Inferior oblique underaction/palsy with subsequent superior oblique overaction.
- Lateral rectus underaction.
- Inferior rectus underaction.

Treatment

Without oblique muscle dysfunction (Fig. 16.80):

1. **'A' pattern esotropia** (Fig. 16.81) is treated by bilateral medial rectus recessions and upward transposition.
2. **'A' pattern exotropia** (Fig. 16.82) is treated by bilateral lateral rectus recessions and downward transposition.

Principles of surgery

The aims of surgery on the extraocular muscles are to correct misalignment and, if possible, restore BSV. However, the first step in the management of childhood strabismus involves correction of any significant refractive error and/or amblyopia. Once maximal visual potential is reached in both eyes, any residual deviation can be treated surgically. The three main types of procedures are: (a) *weakening*, which decreases the pull of a muscle, (b) *strengthening*, which enhances the pull of a muscle, and (c) those that *change the direction of muscle action*.

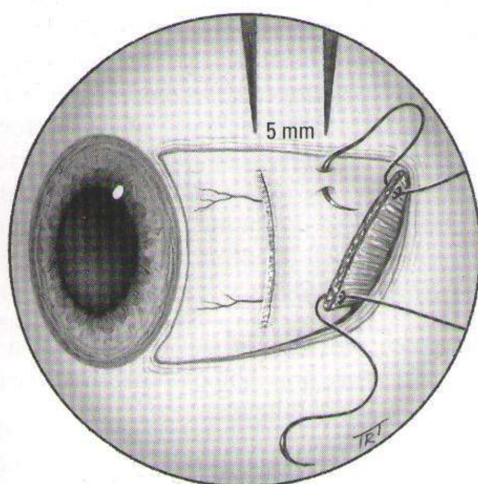


Fig. 16.83
Horizontal rectus muscle recession

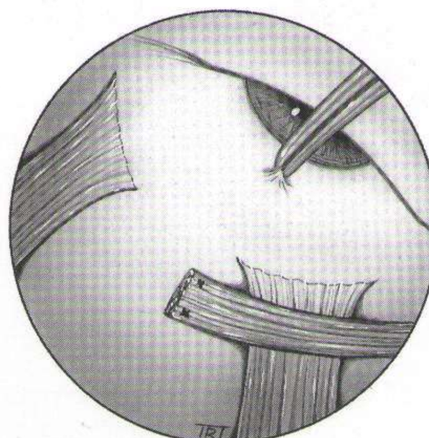


Fig. 16.84
Inferior oblique recession

Weakening procedures

The procedures for weakening the action of a muscle are: (a) *recession*, (b) *myectomy* and (c) *posterior fixation suture*.

Recession

This slackens a muscle by moving its insertion towards its origin. It can be performed on any muscle except the superior oblique.

1. **Rectus muscle recession**
   a. The muscle is exposed and two absorbable sutures passed through the outer quarters of the tendon.
   b. The tendon is disinserted from the sclera, and the amount of recession is measured and marked on the sclera with calipers.
   c. The stump is resutured to the sclera posterior to its original insertion (Fig. 16.83).
2. **Inferior oblique recession**
   a. The muscle belly is exposed through an inferotemporal fornix incision.
   b. One or two absorbable sutures are passed through the muscle near its insertion.
   c. The muscle is disinserted and the stump resutured to the sclera, 2 mm posterior and temporal to the temporal edge of the inferior rectus insertion (Fig. 16.84).

Myectomy

This involves severing the muscle from its insertion without reattachment. It is most commonly used in weakening an overacting inferior oblique muscle. Very occasionally, the procedure is performed on a severely contracted rectus muscle.

Posterior fixation suture

The principle of this (Faden) procedure is to decrease the pull of the muscle in its field of action without affecting it in the primary position. The Faden procedure may be used to treat DVD, although it can also be used to weaken a horizontal rectus muscle. When treating DVD, the superior rectus muscle is usually first recessed. The belly of the muscle is then anchored to the sclera with a non-absorbable suture about 12 mm behind its insertion.

Strengthening procedures

1. **Resection** shortens a muscle to enhance its effective pull. It is suitable only for a rectus muscle and involves the following steps:

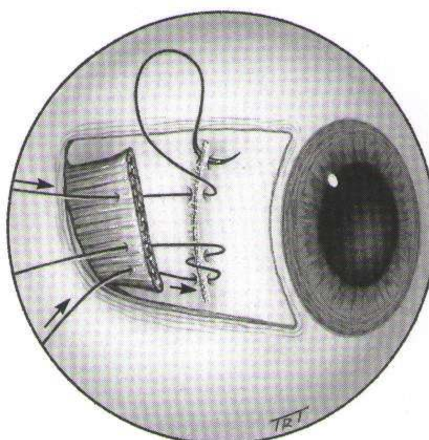


Fig. 16.85
Horizontal rectus muscle resection

a. The muscle is exposed and two absorbable sutures are inserted into the muscle at a predetermined point behind its insertion.
b. The muscle anterior to the sutures is excised and the stump is reattached to the original insertion (Fig. 16.85).

2. **Tucking** of a muscle or its tendon is usually reserved to enhance the action of the superior oblique muscle in congenital fourth nerve palsy.
3. **Advancement** of the muscle nearer to the limbus can be used to enhance the action of a previously recessed rectus muscle.

Treatment of paretic strabismus

Lateral rectus palsy

Surgical intervention for a sixth nerve palsy should be considered only when it is apparent that spontaneous improvement will not occur. This is usually after 6 months have elapsed. The two main procedures aimed at improving abduction are the following:

1. **Hummelsheim procedure**
   a. The medial rectus is recessed.
   b. The lateral halves of the superior and inferior recti are disinserted and reattached to the superior and inferior margins of the paretic lateral rectus muscle (Fig. 16.86a).

> ***NB:*** As this procedure involves detaching three rectus muscles from the globe, there is a risk of postoperative anterior segment ischaemia. To avoid this complication, the medial rectus recession may be replaced with botulinum toxin chemodenervation (see later).

2. **Jensen procedure** is also used to improve abduction and is combined with recession of or injection of botulinum toxin into the medial rectus muscle.
   a. The superior, lateral and inferior recti are split lengthways.
   b. With a non-absorbable suture, the lateral half of the superior rectus is tied to the superior half of the lateral rectus, and the inferior half of the lateral rectus is tied to the lateral half of the inferior rectus (Fig. 16.86b).

Superior oblique palsy

Surgical intervention should be considered in cases of abnormal head posture and diplopia unresponsive to prisms. General principles are as follows:

1. **Congenital** cases with a large hypertropia in the primary position are treated by superior oblique tucking.
2. **Acquired**
   a. Small hypertropias are treated by ipsilateral inferior oblique weakening.
   b. Moderate to large acquired hypertropias may be treated by ipsilateral inferior oblique weakening combined with ipsilateral superior rectus weakening and/or contralateral inferior rectus weakening. It should be noted that weakening the inferior oblique and superior rectus of the same eye may result in defective elevation.
   c. Pure excyclotropias, without hypertropia, are treated by the Harada–Ito procedure, which involves splitting and anterolateral transposition of the lateral half of the superior oblique tendon (Fig. 16.87).

Adjustable sutures

Indications

In certain cases, the results of strabismus surgery can be improved by the use of adjustable suture techniques. These

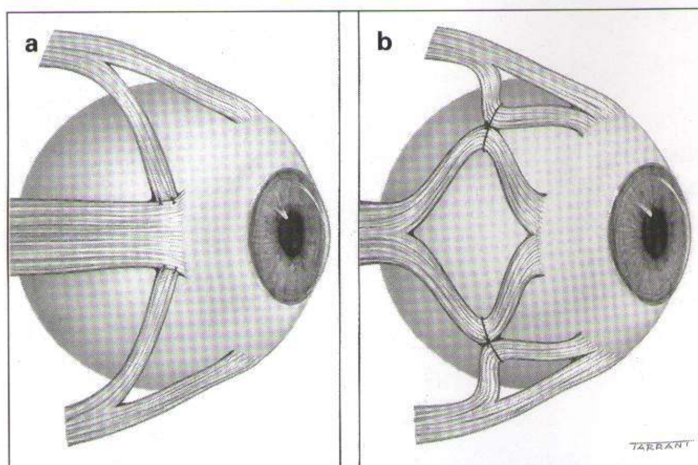


Fig. 16.86
Treatment of lateral rectus palsy. (**a**) Hummelsheim procedure; (**b**) Jensen procedure

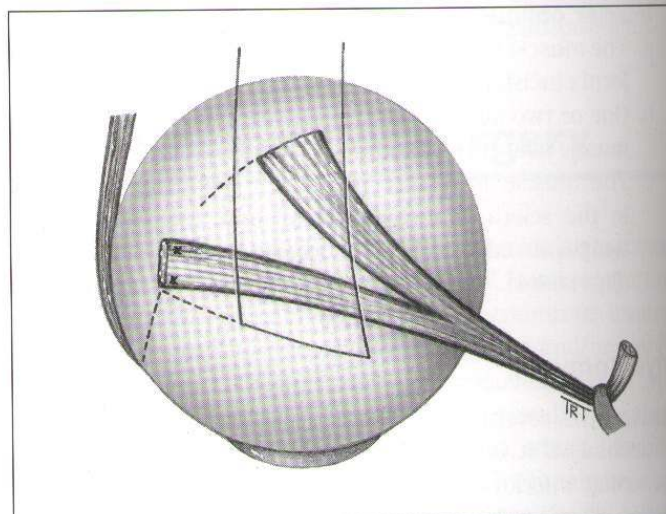


Fig. 16.87
Harada–Ito procedure for superior oblique palsy

are particularly indicated when a precise outcome is essential and when the results with more conventional procedures are likely to be unpredictable; for example, acquired vertical deviations associated with thyroid myopathy or following a blow-out fracture of the floor of the orbit. Other relative indications include sixth nerve palsy, adult exotropia and reoperations in which scarring of surrounding tissues may make the final outcome unpredictable. The main contraindication is a patient who is too young or unwilling to cooperate during postoperative suture adjustment.

Initial steps

1. The muscle is exposed, sutures inserted and the tendon disinserted from the sclera (as for a rectus muscle recession).
2. The two ends of the suture are passed, close together, through the stump of the insertion.
3. A second suture is knotted and tied tightly around the muscle suture anterior to its emergence from the stump (Fig. 16.88a).
4. One end of this suture is then cut short and the two ends tied together to form a loop (Fig. 16.88b).
5. The conjunctiva is left open.

Postoperative adjustment

1. The extent of alignment is assessed.
2. If ocular alignment is satisfactory the muscle suture is tied and its long ends cut short.
3. If more recession is required, the knot is pulled anteriorly along the muscle suture, thereby providing additional slack to the recessed muscle and enabling it to move posteriorly (Fig. 16.88c).
4. If less recession is required, the muscle suture is pulled anteriorly and the knot pulled against the muscle stump (Fig. 16.88d).
5. The conjunctiva is closed.

A similar technique is used for rectus muscle resection.

Botulinum toxin chemodenervation

Temporary paralysis of an extraocular muscle can be created in conjunction with the transposition procedures as already described or in isolation. The following are the main indications for considering chemodenervation:

- To determine function of the lateral rectus in a sixth nerve palsy where medial rectus contracture prevents any abduction. A tiny dose of botulinum toxin B is injected into

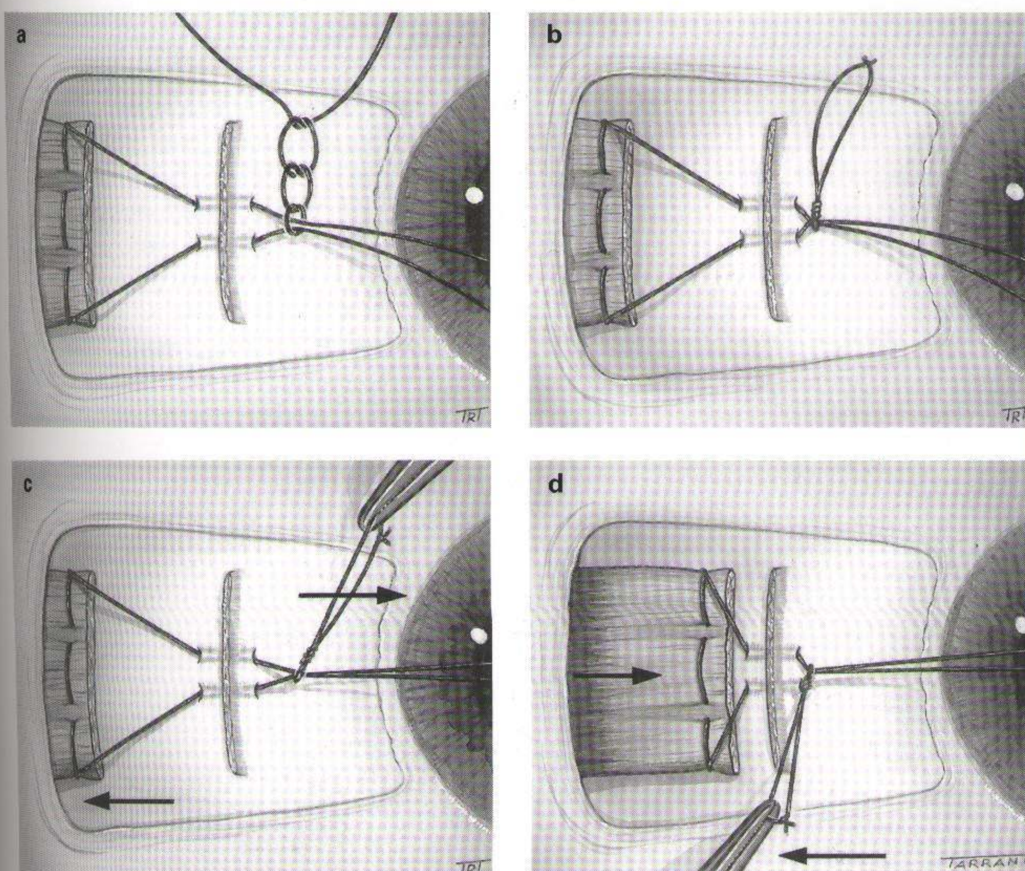


Fig. 16.88
Adjustable sutures (*see text*)

the belly of the overacting antagonist (medial rectus) under electromyographic guidance (Fig. 16.89a). The temporary paralysis of the muscle causes relaxation so that the horizontal forces on the globe are more balanced, thus allowing assessment of lateral rectus function (Fig. 16.89b).

- To determine the risk of postoperative diplopia and to assess the potential for BSV. For example, in an adult with a left divergent strabismus and good vision in both eyes, the left lateral rectus muscle can be injected so that the eyes will either straighten or converge.
- However, placing a corrective prism in front of the deviating eye offers a simpler, and perhaps more accurate method of determining the risk of postoperative diplopia.

If either of the above indicates the likelihood of diplopia, the patient can then be advised preoperatively of this possibility. Such diplopia, however, almost invariably resolves spontaneously.

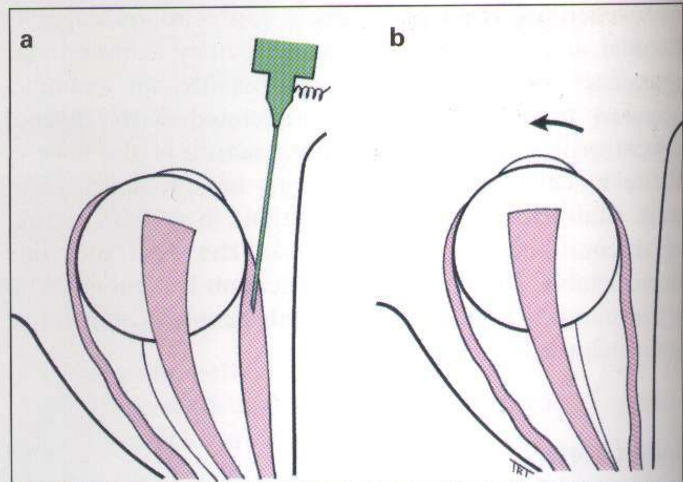


Fig. 16.89
Principles of botulinum toxin chemodenervation in left sixth nerve palsy (*see text*)